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A CASE OF ACUTE BULBAR PARALYSIS,

WITH REMARKS ON ITS PATHOLOGY AND DIAGNOSIS.

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A CASE OF ACUTE BULBAR PARALYSIS,

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The number of contributions to medical literature on the subject of acute bulbar paralysis is a limited one. For our knowledge of its pathological anatomy and symptomatology we are indebted chiefly to the descriptions given by Leyden, Hallopeau, Lichtheim, Erb, Schwalbe, Senator, Hughlings Jackson, Oppenheim, Gowers, and Etter, whose cases have, for the most part, afforded an opportunity of post-mortem examination. In the majority of instances the pathological changes had their origin in the blood-vessels which had become obliterated by thrombosis or, exceptionally, by embolism. Multiple foci of softening, of varying extent, were most frequently found. In rarer cases hæmorrhages (from vascular disease or traumatism), compression by tumor, or an acute or subacute polioencephalitis inferior, constituted the lesions that gave rise to the paralysis. The gravity of lesions of the medulla oblongata is easily understood when it is remembered that within a narrow space it not only gives passage to the fibres that connect the brain with the spinal cord, but that it also contains the centres

of most of the cranial nerves, and especially those which control the cardiac and respiratory functions.

The case I am about to describe is of considerable clinical interest, presenting, as it does, a well-defined and practically typical picture of acute unilateral bulbar paralysis. It demonstrates, in addition, the efficacy of treatment with the iodides which I regard as no less valuable (in smaller doses) in angeio-sclerosis than in syphilitic endarteritis and the sclerotic changes following it.

Mr. J. S. D., thirty-nine years of age, unmarried, vocal teacher, was referred to me on the 17th of last October, and gave the following history:

Both parents, as well as seventeen brothers and sisters, are still living and in good health. He had measles, whooping-cough, and varioloid in his childhood. In 1877 he had a sunstroke, in 1886 rheumatic gout, and in 1889 a mild attack of influenza. For the greater part of his life, up to three years ago, he has suffered from what he calls bilious headaches. His occupation has never exposed him to mental or physical overwork, but his habits have, he admits, not infrequently led to alimentary excesses, especially during his operatic career. He has always used stimulants very moderately, however, and has never been addicted to the use of tobacco. Denies ever having been infected with syphilis.

Three weeks before coming under my observation he was seized during the night with a dull pain in the back of the head and neck, with dizziness and ringing in the right ear. Dimness of vision, together with an indisposition to mental and bodily exertion, had been coming on, he thinks, for three or four months. He found that the dizziness disappeared when he was in the recumbent posture, but that his whole body swayed sideways when he turned over on the left side. Three days later he noticed some embarrassment in swallowing and a numbness of the right foot.

These troubles continued unchanged until the morning of the tenth day following, when he awoke with a numbness

and tingling in the right half of his face and in the right arm and leg. The lips and tongue felt stiff, and the difficulty in swallowing had become more pronounced. His discomfort was increased by difficulty in speech, shortness of breath, and by seeing objects double. He also noticed that his head was inclined somewhat to one side, and that the back of his neck, the right arm and leg were weak. Walking had become so unsteady that he was unable to get about unless he was supported. For a day or two his mind was somewhat confused, so that he did not remember clearly afterward what he had said or done during that time. Nausea or vomiting has at no time been present, and there has been no fever. Sleep has been poor since he was taken ill.

Status Præsens.—Patient is somewhat under medium height, fairly well nourished, of active, nervous temperament, and looks at least ten years older than he is. The radial and temporal arteries are rigid, the latter markedly serpentine. There exists some enlargement of the præcordial area of dullness, and the second sound is accentuated, but otherwise no cardiac abnormity can be made out. The pulse is not altered in frequency or rhythm. On the part of the lungs and abdominal organs no morbid signs are to be detected, and evidences of constitutional syphilis are wanting. The sphincters are intact. The urine is normal in quantity and contains neither albumin nor sugar. The existing amblyopia does not appear to depend upon any changes of the optic papilla which an ophthalmoscopic examination shows to be in normal condition. The cephalalgia is occipital, extending down the neck, and not subject to exacerbation; it has the character of a dull ache. Psychically, he presents nothing unusual beyond a great depression and an inclination to weep at the slightest provocation.

The head is slightly inclined to the left. When the patient is standing or sitting it falls forward, and in order to straighten it he has to resort to the use of his hands. The turning of the head to either side is not impeded, however, and passive movements are readily executed. The vertigo is increased by turning it to the left.

The movements of the right external rectus are weak, and efforts to turn the eye outward give rise to nystagmic twitching. Diplopia is marked. The pupils are equal and moderately contracted; they react to light and accommodation. The frontal muscles contract symmetrically, but, owing to weakness of the orbicularis palpebrarum, the right eye can not be closed as firmly as the left; the right naso-labial fold is somewhat effaced; the corner of the mouth, slightly opened, hangs down a little on the same side and permits a constant dribbling of saliva, especially when he is lying down. The soft palate appears lower on the right and slightly drawn toward the opposite side, but the uvula is not deflected.

Owing to paralysis of the right half of the tongue, deglutition and speech are interfered with. Food can be moved about in the mouth only with difficulty, and articulation is indistinct, particularly as regards the pronunciation of the labials and linguals. It is only with the aid of a draught of water or by pushing it backward with the finger that he is able to transfer his food to the pharynx. When at rest within the buccal cavity the tongue shows nothing abnormal, but deviates to the right when protruded. The voice is somewhat raucous and dyspnœa is still complained of. The muscles supplied by the motor portion of the fifth nerve are intact.

There exists a feebleness of the extremities of the right side. Muscular exertion, even the most moderate, is followed by great fatigue which can not be wholly attributed to the dyspnœa from which he is suffering. No tremor, either of the tongue or extremities, is present.

The sensory disturbances consist in numbness and tingling of the right half of the face and corresponding mucous membranes, as well as of the right arm and leg. Objectively, no alteration of sensation is discoverable beyond a trivial hypæsthesia on the same side. Except in the right half of the tongue, the tactile sense is not affected, and there is no dissociation of sensibility.

The sense of taste is impaired in the right half of the tongue, the patient being unable to distinguish with certainty sweet, bitter, and acid substances. The soft palate, tonsil,

and upper pharynx are somewhat anæsthetic on the right side. Hearing is diminished in the right ear, but the sense of smell is not affected.

The gait is titubating, with a tendency to stagger toward the right. It is so marked that walking is impossible without assistance. When he is asked to stand with his eyes closed, the ataxia is so great as to cause him to fall (to the right). In order to avoid staggering he enlarges his basis of sustentation by widely separating his feet and keeps a fixed point in view in front of him. Being conscious of the extent and direction of the movements he executes in walking, he does not watch his feet as he would do were the ataxia of the locomotor type. The inco-ordination does not extend to the upper extremities.

The cutaneous and tendon reflexes are not altered; there is no exaggeration of the masseteric reflex.

Under the use of iodide of potassium, appropriate diet, and galvanization, at a later period, of the nucha and through the mastoid processes, as advocated by Strümpell and Oppenheim, the symptoms have gradually subsided, so that now, five months since the beginning of treatment, only slight numbness and tingling of the tips of the fingers of the right hand remain. The vision has been completely restored and his lacrymose frame of mind has given place to one of comparative impassibility.

The varying distribution of the vascular supply of the medulla oblongata has made it impossible so far to separate into syndromal groups the disturbances that arise from arterial obstruction. But the frequent anomalies of arterial distribution do not form the only obstacle to such a separation; it is largely upon the multiplicity and situation of the points of obstruction and the shape, which is often irregular, of the necrobiotic (or hæmorrhagic) foci that the symptomatology depends. That mere narrowing of the lumen of a vessel, altered by atheromatous or endarteritic processes, may, by retarding the blood current and favoring

coagulative adhesions, be the cause of morbid symptoms is readily apparent. The possibility of compression by rigid, dilated, and frequently tortuous sclerotic vessels is another factor to be taken into account.

According to Charpy, the floor of the fourth ventricle derives its blood supply from two different sources: the upper half, which contains the nuclei of the facial, abducens, and trigeminus, receives its nutrient vessels from the basilar artery; the lower half is supplied by the ventricular branches of the anterior and posterior spinal arteries which are given off by the vertebral. An occlusion of the basilar would therefore, when not compensated by collateral circulation, give rise to bilateral lesions, whereas an obliteration of one of the vertebral arteries would be productive of unilateral lesions.

Duret, whose elaborate researches on this subject, published in 1873, have in the main been confirmed by those of Adamkiewicz, divides the vessels of the medulla oblongata —all of them branches of the vertebral artery—into (1) the radicular arteries, which divide into two branches—one accompanying the nerve root to the periphery, the other following it to its nucleus; (2) the median (or nuclear) arteries, of which some, coming from the anterior spinal, supply the nuclei of the spinal accessory, hypoglossal, and inferior facial, while others supply the pneumogastric, glossopharyngeal, and auditory nuclei, and others again, given off by the basilar, pass across the fibres of the pons to reach the nuclei of the superior facial, abducens, patheticus, and motor oculi; (3) arteries of the remaining portions of the medulla oblongata, which take their origin from the vertebral, anterior spinal, and inferior cerebellar arteries, and are distributed to the olivary bodies, pyramids, lateral tracts, and corpora restiformia. The last two receive their supply from the inferior cerebellar branch.

How are we to interpret the complexus of symptoms I have described? The mode of onset and character of the disease, the evidences of arterio sclerosis presented by the patient, and the steady improvement under treatment jus-

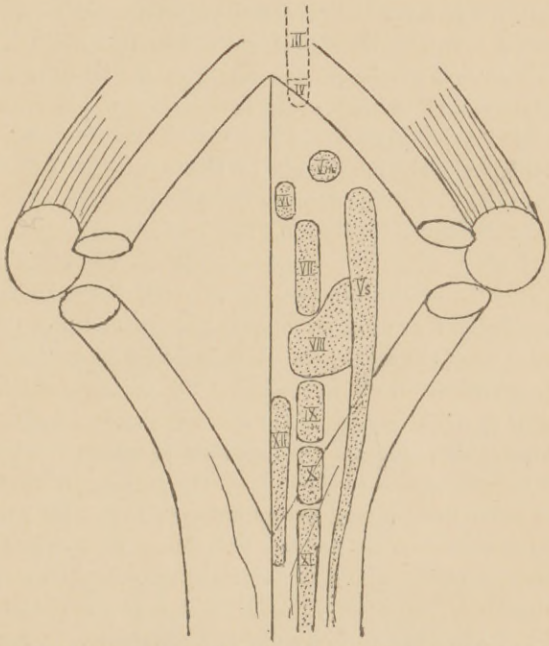

Diagram showing arrangement of nuclei in floor of fourth ventricle (right side). III, motor oculi; IV, patheticus; Vm, motor nucleus of trigeminus (locus cæruleus); Vs, sensory (descending) root of trigeminus; VI, abducens; VII, facial; VIII, auditory; IX, glosso-pharyngeus; X, vagus; XI, spinal accessory; XII, hypoglossus.

tify the conclusion that it was caused by thrombotic obstruction of the right vertebral artery. It would account for the involvement, more or less pronounced, of the hypoglossal, vago-accessory, glosso-pharyngeal, auditory, abdu-

cens, facial, and sensory trigeminus, as well as for the hemiparesis and hemiparæsthesia of the right side. The accompanying diagram will serve to recall the arrangement of the nuclei in the floor of the fourth ventricle. It is safe to conclude, from the absence of any appreciable source of embolism and from the existence of prodromal symptoms for some length of time, that the obstruction was not due to an embolus, which, besides, rarely passes into the vertebral artery, and in that event is more apt to plug the left branch.

It is obviously impossible to locate with accuracy the seat of the lesion that led to the right hemiparesis. Whether, in unilateral affections of the medulla oblongata, the hemiplegia is direct or alternate depends not alone upon the situation of the diseased focus relative to the motor decussation—*i. e.*, upon its situation above or below this—but also upon the seat of the obstruction (in cases of thrombosis or embolism) above or below the point at which the anterior spinal artery is given off. The direct hemiparesis may have been due in this instance to an encroachment upon the motor fibres below their decussation, or, what appears to me more probable, to a lesion of the direct pyramidal tract above that point. The latter view seems the more acceptable in the light of the recent discovery of Jacobsohn (*Neurol. Ctrlbl.*, 1895, No. 8, p. 348), who has found that the fibres of the direct pyramidal tract are not intermingled with those of the crossed pyramidal tract, but that in their entire course through the medulla oblongata they occupy the lateral angle of the pyramids. There is no satisfactory evidence showing that they cross at successive levels of the spinal cord and enter the anterior cornua of the opposite side; such a conception has always seemed to me, on physiological grounds at least, open to doubt. Competent observers have of late called it into

question, and advanced the opinion that the fibres terminated in the gray matter of the same side. If, as has been believed hitherto, they do not reach below the middle or lower dorsal region, my conjecture would leave the paresis of the lower extremity still unexplained. Quite recently, however, at a meeting of the *Société de biologie*, Déjerine and Thomas (*Bulletin médical*, 1896, No. 13; *Médecine moderne*, 1896, No. 13) have been able to demonstrate, by a series of sections treated according to Marchi's method, that the column of Türck can be traced as far as the root of the fourth sacral nerve. From the lower dorsal region downward it appeared in the sections no longer in the shape of a comma, but is a small mass in front of the anterior commissure.

How great the difficulty in accounting for unilateral paralysis or anæsthesia in pontine or bulbar affections can be is shown by the case of Langdon (*Brain*, 1895, lxxii), in which compression by a tumor (fibro-cystoma of pons and cerebellum) above both the motor and sensory decussations produced motor and sensory disturbances on the same side. The author attempts to account for them by assuming that they were "due to a *contre-coup* effect—*i. e.*, to pressure by the tumor of the right periphery of the medulla against the comparatively acute margin of the foramen magnum at a point above (before) the motor decussation and also above (after) the sensory (fillet) decussation." Another case in point is that reported by Senator (*Arch. f. Psych. u. Nervenkrh.*, 1882, xi, p. 713), in which a focus of softening in the left half of the medulla oblongata, one centimetre in its longest diameter, caused, among other symptoms, crossed hemianæsthesia affecting the left face and right extremities.

Could the syndrome observed in the patient have originated in an extrabulbar lesion, such as an aneurysm of the vertebral artery or a neoplasm? A growth of this

kind, when it compresses the cranial nerves and the pyramidal tract of the same side below its decussation, may produce peripheral paralysis of the cranial nerves with hemiplegia on the same side, but, in view of the clinical history, especially the acute development of the headache and the absence of vomiting, it is unlikely that this was the case. Moreover, the prompt results of the treatment speak against the existence of an aneurysm or any other form of tumor, except a gumma, which would, however, have required far more energetic medication than my patient has been subjected to.

A striking feature of the case was the extreme incoordination. As a result of his experiments on the frog, Foster, in his *Text-book of Physiology*, long ago expressed the opinion that in the mammal as well as in the frog the medulla oblongata was largely concerned in the co-ordination of movements. Anatomico-clinical facts have since made it evident that it contains a region whose lesion produces disturbance of equilibration. It is probable that this region is to be found in the bulbar endings of the posterior columns of the spinal cord. Oppenheim (*Lehrbuch der Nervenkrankheiten*, 1894, p. 606) is inclined to place it in the interolivary tract, and perhaps in the corpus restiforme. Ataxia of the "cerebellar type" has been observed in connection with a lesion of the corpus olivare. The case of Senator's, previously cited, in which a focus of softening was found in the left half of the medulla oblongata, showed a marked tendency to fall to the left. Dinkler (*Deutsche Zeitschr. f. Nervenkrh.*, vii, 1895, p. 465) has reported a case of acute hæmorrhagic polioencephalitis inferior, the result of cranial traumatism, in which, in addition to a train of symptoms indicative of bulbar involvement, there were a staggering gait and an occasional tendency to fall. The patient succumbed two years and a half

after he had received the injury, and the autopsy revealed numerous small extravasations in the floor of the fourth ventricle, particularly noticeable in the nuclei of the fifth, eighth, and tenth nerves, and a cellular infiltration of the walls of the blood-vessels. Hæmorrhages existed also in the posterior cornua of the cervical spinal cord, but with the exception of a diffuse cerebro-meningeal hyperæmia no other lesions were found in the nerve centers. It is interesting to note in this connection that several authors, like Luciani, Dupuy, Eisenlohr, Bristowe, and Bruns, contrary to the view generally held, do not regard inco-ordination as pathognomonic of disease of the cerebellum, whether this involves the vermis or not. Their experiences are in accord with those of Friedeberg (*Berliner klin. Wochenschr.*, 1895, No. 33), who found that, out of nine cases of cerebellar disease the diagnosis of which was verified by the autopsy, only two showed an ataxic gait.

While a paresis of the right sterno cleido mastoid and trapezius muscles—the latter in so far as its clavicular portion, innervated by the spinal accessory, is concerned—would explain the oblique position of the head, it is not sufficient to account for the weakness of the posterior cervical muscles which, next to the difficult deglutition, dysarthria, and inco-ordination, seemed to the patient the most formidable symptom. There was no evidence that the drooping of the head was due to an involvement of both trapezii.

Together with the proneness to general muscular exhaustion, it recalls the form of bulbar paralysis without anatomical lesions, which has been studied by a number of observers, chief among them Wilks, Erb, Oppenheim, Shaw, Goldflam, Hoppe, and, more recently, by Pineles, Jolly, Murri, and Strümpell, of whom the last named has published an exhaustive paper treating of this topic a short

time since (*Deutsche Zeitschr. f. Nervenhkr.*, 1896, viii, p. 16). This progressive and habitually fatal disease is characterized by an abnormal tendency to muscular fatigue, and has for this reason been termed asthenic bulbar paralysis by Strümpell and myasthenia gravis pseudo-paralytica by Jolly. The paresis affects not only the muscles of mastication and deglutition, thereby producing dysphagia and dysarthria, but also those of the trunk, extremities, and notably of the back of the neck. Ptosis, diplopia, and disturbed innervation of the upper and lower facial nerves have been the rule in the cases on record, while sensory and psychical disorders have been absent or very slight.

The absence of fibrillary tremor and atrophy, the presence of sensory disorders, and the fact that the symptoms attain their maximum within a very short period of time after their inception, sufficiently distinguish, I think, acute (bilateral) bulbar paralysis from the labio-glosso laryngeal paralysis of Duchenne. Its differentiation, however, from the labio-glosso-laryngeal paralysis of cerebral origin, or pseudo-bulbar paralysis, may be attended with difficulty. Before the bulbar symptoms make their appearance in the latter affection, repeated apoplectic attacks have generally occurred and, as a rule, the lower extremities have been found to be more affected than the upper, in which there is often a mere motor weakness. In two cases I had an opportunity of observing several months ago in Professor Raymond's clinic at the Salpêtrière a rapid amelioration of the hemiplegic symptoms took place, while the bulbar phenomena remained unimproved. This is, indeed, the usual course of the malady, although cases have been reported in which the apoplectic seizure and hemiplegia were wanting, the bulbar symptoms appearing *d'emblée.*

Pseudo-bulbar paralysis depends, in the large majority

of cases, upon symmetrical lesions of both cerebral hemispheres, the foci of softening being seldom cortical, but mostly in or about the basal ganglia, notably in the external portion of the lenticular nucleus, the putamen (Grasset et Rauzier, *Maladies du système nerveux*, 1894, ii, p. 26). They have also been found in the centrum ovale, cerebellum, and pons. Unlike acute bulbar paralysis, in which the psychical manifestations are limited to a tendency to weeping or, less often, to laughing (Oppenheim), it is always associated with impairment of the intelligence, with varying degrees of dementia, confusion, excitability, or loss of memory. The general sensation as well as the special senses are almost always intact, although Oppenheim and Siemerling have noted optic changes. Cephalalgia, vertigo, disorders of the ocular muscles, and cardio-pulmonary troubles are absent, and laryngeal symptoms seem to be exceptional.

When ushered in by an apoplectoid ictus, the bulbar form of disseminated sclerosis may assume the character of an acute bulbar paralysis, but a consideration of the patient's age, the mode of evolution, and the presence of intention tremor will be sufficient to make the differential diagnosis. Ataxia occurs in this disease only when it is complicated with tabes.

In the course of syringomyelia localized in the medulla oblongata, of which the chief symptoms are troubles of speech and deglutition, apoplectoid attacks are likewise not infrequent, and the disease may for this reason bear a close resemblance to acute bulbar paralysis. The further progress of the syringomyelia, its characteristic sensory disorders, and the muscular atrophies will remove the doubt.

As regards the diagnosis from tabes dorsalis, a confusion is scarcely possible even when, as is sometimes the

case, the vago-accessorius and sensory trigeminus are implicated and vertigo exists. The same may be said of those varieties of amyotrophic lateral sclerosis and acute ascending paralysis (Landry's paralysis) which begin with bulbar symptoms.

It is well to bear in mind, finally, that there is hardly an affection of the nervous system, acute bulbar paralysis not excepted, which hysteria may not simulate.

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