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DEAFNESS DUE TO LESIONS IN THE BRAIN

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Total bilateral deafness due to lesions in the brain appears to be an exceedingly rare symptom, for very few cases are to be found in the literature of neurology in which it has been present. It is well known that the cortical centers of hearing lying in the temporal convolutions receive impulses from both ears and hence a lesion in one hemisphere is not necessarily attended by total deafness. A few cases have been recorded in which lesions in both temporal lobes have occurred and have resulted in a state of total deafness, but in all these cases (and they are not many) the lesions in the two hemispheres have not occurred simultaneously, and hence the patient has not been suddenly afflicted with total deafness. There is, however, a sufficient number of cases on record with autopsies to prove that when both temporal lobes are destroyed by disease a patient will necessarily be totally deaf.

The connection between the temporal lobes and the termination of the acoustic nerves in the medulla and pons is now well known. The primary centers in which the acoustic fibers end lie at the junction of the medulla with the pons and consist of several groups of cells lying partly upon the floor of the fourth ventricle and partly deep within the formatio reticularis. From these centers tracts arise which pass upward to the cortex, forming the central acoustic tract. Like all sensory tracts the acoustic tract is not a continuous one, but is made up of a series of

short fibers with intercalated neurones. The chief decussation of this tract occurs low down in the pons in the trapezoid body, but this decussation is not a complete one and hence each acoustic tract conveys impulses from both ears. That this tract is not a continuous one from the nuclei of origin to the cortex may be deduced from the very large number of reflex acts which any auditory impulse will produce. A loud sound upon one side causes an involuntary start, a turning of the head toward that side; a turning of the eyes toward that side, and not infrequently a sudden dilatation of the pupils and, in addition to these reflex acts, processes of thought are arrested and attention is fixed upon the sound. It is thus evident that the auditory impression is capable of setting up a large number of automatic acts as well as of exciting cortical activity; a fact which implies an interrupted and not a continuous tract.

Investigations of Held,¹ Thomas² and Van Gehuchten³ have shown that the course of this central acoustic tract is chiefly in the lemniscus, or fillet, in its lateral portion and that it has connections with the upper olive, with the nucleus of the lemniscus, with the corpus quadrigeminum posterior, with the corpus geniculatum internum and with the temporal convolutions.

Such being the course of the acoustic tract, it is to be expected that lesions in the pons varolii and in the tegmentum of the crura cerebri which affect the lemniscus should produce deafness, but cases of this kind appear to be extremely rare. For this reason the following case is recorded:

CASE OF AN APOPLECTIC ATTACK CHARACTERIZED BY ALTERNATING PARALYSIS OF MOTION AND OF SENSATION, BY DYS-
ARTHRIA, AND BY TOTAL DEAFNESS IN BOTH EARS.

The patient, whose arteries had become markedly atheromatous, from a long-continued abuse of alcohol, had a slight attack of right hemiplegia and aphasia in the year 1900, when 42 years old. The symptoms rapidly subsided and as the attack was not attended by loss of consciousness, it seemed probable that it was due to a thrombosis in a small vessel, leaving a focus of sclerosis of small extent and without permanent effects. In June, 1901, a second apoplectic attack, however, occurred, more severe in

¹ Held, Arch. f. Anat. und Phys., 1893.

² Thomas, Comptes rend. de la Soc. de Biol., 1898, p. 183.

³ Van Gehuchten, Le Nevraxe, IV, 280, VIII, 58 and 127.

character, the permanent symptoms of which remained until her death in 1909. In this attack she became unconscious and remained so for two days, after which time it was apparent that the right side of her face was paralyzed and that the left arm and leg were paralyzed. The face was flat and it was drawn toward the left side. The right eye could not be firmly closed. The right side of the face was wholly expressionless and saliva drouled from the right corner of the mouth; the tongue was paralyzed and protruded toward the left; the left arm and leg were quite paretic. Sensation in the right side of the face was markedly impaired to touch, temperature and pain, and the same loss of sensation was apparent in the left arm and left leg and upon the left side of the body as high as the collar. The right extremities were also slightly weaker than before the attack.

In addition to the alternating paralysis and anesthesia, there was a very marked loss of muscular sense and this was present in all four extremities, being more marked on the left side. The patient was unable to swallow and choked at any attempt, so that she was fed for a month with difficulty. She was entirely incapable of pronouncing words distinctly and as time went on this difficulty increased, being intensified by her total deafness.

From the day of the attack until her death *she was totally deaf in both ears*. The deafness was complete for all sounds, high or low, and there was no bone conduction of sound. A very loud jangling bell rung behind her caused a sense of uneasiness so that she would move her body restlessly and become aware that something disagreeable was happening. But this was evidently felt as a vibration, because it was not referred to the ears and was not heard. There was no disease of the ears, as determined by Dr. Gorham Bacon. This condition was present for eight years. There was apparently no return of hearing whatever. Articulation was imperfect and it was difficult to understand her as she was unable to enunciate with any clearness.

When last seen in July, 1909, the right side of the face was flat in its lower part, especially when she was fatigued, but the eyes closed normally. She constantly drouled in the right side of the mouth. The tongue protruded straight; swallowing was quite easy, though occasionally she had fits of choking. There was marked paresis of the left arm and leg, the left arm being constantly in a state of contracture, flexion of the elbow, of the

wrist, and of the fingers being present. The leg was in a state of contracture, extended, the foot and toes being markedly extended, but the great toe being constantly dorsally flexed. There was exaggeration of all reflexes; Babinski and Oppenheim reflexes marked on left side. The condition of alternating anesthesia to touch, temperature and pain remained until her death.

There was very marked ataxia in both legs. She was unable to stand alone and any attempt at walking resulted in the most extreme ataxic movements of the legs, more marked upon the left side than upon the right. The legs were thrown in different directions, she did not know where they were and could not guide them unless she watched them and then the guidance was very imperfect. There was considerable ataxia of both hands, much more marked in the left than in the right. She could not perceive accurately, or reproduce positions given to the left fingers.

She was perfectly intelligent, was able to write, and had been able to write from the time of the attack, showing that the condition of disturbance in speech was one of dysarthria and not of aphasia. She read, and understood everything conveyed to her in writing, and by the deaf and dumb finger speech. She was consistent in all her statements. She was in no sense hysterical, and a careful daily observation during four months and occasional observation during eight years convinced me that there was no element of hysteria in the condition of deafness. She died suddenly in a uremic convulsion. No autopsy was allowed.

Diagnosis.—The syndrome of alternating paralysis and anesthesia is characteristic of a lesion in the caudad portion of the pons, in one lateral portion. In this case it must have been upon the right side. The lesion must have affected the pyramidal motor tract in the ventral part of the pons to cause the hemiplegia. It must also have affected both the lemniscus and the formatio reticularis in the dorsal part of the pons to produce the ataxia and the alternating anesthesia. I showed in 1884⁴ that the tract of muscular sense passes in the lemniscus and that the tracts of tactile sense, pain and temperature senses pass in the formatio reticularis. The condition of ataxia present in this patient was similar to that recorded in the cases of Senator,⁵

⁴ JOUR. OF NERV. AND MENT. DIS., July, 1884.

⁵ Archiv f. Psych., XIV, 2.

Kahler⁶ and Pick and Spitzka⁷ in which cases the lesion was limited to the lemniscus. But in my patient the ataxia affected to some extent both legs, indicating that the lemniscus was affected on both sides of the median line. The lemniscus lies near to the deeper part of the pons behind the motor tract.

Dysphagia and anarthria are due to lesions in this locality. Markowski,⁸ in a careful study of the symptoms of anarthria due to pons lesions, attempts to separate sharply between lesions in the ventral and lesions in the dorsal part of the pons, the former causing chiefly motor symptoms of a hemiplegic, unilateral or alternating nature, or possibly bilateral hemiplegia, as in his case. He affirms that dysphagia is due to lesions in the median and dorsal part of both pyramidal tracts in the pons, the speech tract being presumably bilateral, because left unilateral lesions destroying the pyramidal tract do not always cause anarthria, while if such a lesion is present and a second lesion occurs in the right side (as in his case) anarthria appears. The same course is taken by the tract which controls swallowing. For while swallowing is largely reflex and is possible so long as the medullary nuclei are intact, it is usually initiated by a voluntary impulse, and if the tract conveying this impulse is affected, difficulty in beginning the act appears. This was the form of dysphagia present in my case. If she attempted to swallow voluntarily she choked. If the mind was diverted and food taken automatically there was no difficulty. His statements being based upon a careful study of the situation of the lesions in his case may be accepted. He cites 18 cases of unilateral pons lesion. In 10 there was no dysarthria. In two of the remaining 8 the record is imperfect. In 6 dysarthria was present, 3 being on the right and 3 on the left side. In 9 cases of bilateral lesion anarthria was present in all. In none of his cases was deafness present.

The pons lesion seems to afford an adequate explanation of the deafness. We have already seen that the auditory tract decussation is in the trapezoid body, its fibers passing across the raphe around the location of the lemnisci. A lesion which affected both lemnisci in my patient could not fail to have involved both auditory tracts at their decussation and hence would arrest impulses from both ears. That the nuclei of the

⁶ Vierteljahrschrift f. Pract. Heilk., Bd. 142, 596.

⁷ Amer. Jour. of Neurology and Psychiatrie, November, 1883.

⁸ Arch. f. Psych., XXIII, p. 381.

acoustic nerve were not affected seems to be indicated by the fact that when a very loud noise was made behind the patient she would move her head reflexly without knowing why and would feel a sense of discomfort. This would seem to show that sensations reaching these nuclei through the nerve caused reflex impulses to the spinal accessory and cervical centers but failed to set up an impulse sufficiently strong to overcome the break in the auditory tract.

The question of the nature of this lesion seems easy to determine. A thrombus in one of the branches of the basilar artery which enter the raphé of the pons and send their branches laterally into its structure, might easily produce an area of softening and sclerosis, affecting the median part of the pons and its right half in the caudad portion.

The following cases have been found in literature to confirm this diagnosis and to prove that a lesion of the pons may cause deafness. As they were all unilateral and did not affect the decussation of the auditory tract the deafness in all these cases was unilateral.

Case 1.—In a case described by Miles⁹ a hemorrhage in the left half of the pons, involving the nuclei of the v, vii and viii nerves and the lemniscus and pyramidal tract, caused alternating paralysis and anesthesia and total deafness in the left ear.

Case 2.—Miles also recorded¹⁰ a case of tumor of the left half of the pons, producing similar symptoms.

Case 3.—In a case described by P. F. Becker¹¹ of an infiltrating glioma of the pons deafness was present. The patient suffered during ten months from progressive symptoms. Vomiting and vertigo first appeared; soon after right vii n. and right vi n. paralysis with optic neuritis followed. Then difficulty in articulation and in swallowing with loss of taste, and paresthesia in the mouth appeared. Then the right v n. was paralyzed with inability to chew. A general weakness of the extremities with ataxia and finally marked anesthesia developed. *The hearing was diminished in both ears, but the right was deafer than the left.* The autopsy showed an infiltrating glioma affecting the right half and median portion of the pons and compressing all the tracts. It projected into the fourth ventricle.

Case 4.—In a case described by Warfvinge¹² of a gelatinous infiltrating tumor of the medulla on the left side there was deaf-

⁹ Archives of Medicine, Aug., 1882.

¹⁰ Archives of Medicine, Oct., 1881.

¹¹ Arch. f. Psych., XXXV, 503.

¹² Neurol. Centralbl., 1889, Vol. VIII, p. 461.

ness in the left ear. The patient suffered from vertigo, vomiting and headache, from paralysis of the left vi and vii nerves, and of the right arm and leg. The tongue protruded to the right.

Case 5.—In a case described by Glaezer¹³ of a tumor in the posterior and lower part of the fourth ventricle which had compressed the pons there was deafness in both ears, especially marked in the right ear.

Case 6.—In a case recorded by Ranschoff of softening in the dorsal part of the pons involving the lemniscus on the left side the motor, ataxic, articulatory symptoms resembled those present in my patient. Unfortunately no mention is made of the condition of hearing although hallucinations of hearing were present.¹⁴

Case 7.—In a case described by v. Bechterew in which the dysarthria resembled that in my patient no tests of hearing are recorded.¹⁵

Case 8.—In a careful study of pons lesions by Dana¹⁶ nine cases are recorded which present typical symptoms of lesions of the pons some being hemorrhages, others foci of softening. In one only of these cases is any record found of the condition of hearing; in that one the patient was slightly deaf in the left ear and in this the lesion was an extensive softening in the left side affecting the lemniscus.

Case 9.—In a case described by Babinski and Nageotte¹⁷ of softening in the interolivary tract and lemniscus on the left side due to thrombosis of branches of the basilar artery the symptoms in many respects resembled those in my patient. There was a marked "hemiasynergie" of the left leg, lateropulsion toward the left; tremor of the upper extremities, right hemiplegia and hemianesthesia, difficulty of deglutition. The patient walked with help only, both feet being markedly ataxic especially the left foot. In this history there is no mention of the condition of hearing but the lesion did not affect the lateral part of the lemniscus in which the auditory tract is supposed to lie.

Case 10.—Moehi and Marinesco¹⁸ have recorded a case of lesion in the dorsal half of the pons, and have collected 15 other cases similar to their own. The symptoms present were permanent paresthesia in the entire left side of the body, diminution of pain sense and temperature sense without total loss of touch, inability to locate the position of the limbs, some ataxia of the left hand, uncertainty in the gait, vertigo, no paralysis of the limbs, left unilateral paralysis of the jaw and slight right sided abducens and facial paralysis which finally disappeared. The lesion lay in the right half of the pons, in its dorsal part,

¹³ Deut. Med. Woch., 1897.

¹⁴ Arch. fur. Psych., XXXV, 413.

¹⁵ Deut. Zeit. für Nervenheilkunde, XVII, 223.

¹⁶ N. Y. Medical Record, Sept. 5, 1903.

¹⁷ Icon. photo. de la Salpêtrière, Vol. XV, p. 492, 1902.

¹⁸ Arch. f. Psych., XXIV, 666.

not reaching the raphe, nor the floor of the ventricle but involving chiefly the formatio reticularis and to a small extent the lemniscus in its median portion opposite the vi n. nucleus. In this case hearing was not affected; there was no dysarthria or dysphagia.

Conclusions.—From a review of these cases it may be concluded that: (1) Deafness may be produced by lesions of the pons varolii; (2) the deafness will be on the side of the lesion if the acoustic nucleus only is affected; (3) the deafness will be bilateral if the trapezoid fibers are involved at their decussation in the raphe; (4) the deafness will be on the side opposite to the lesion if the superior olivary nucleus and the lateral part of the lemniscus are affected in the pons.

As to the effect of lesions in the auditory tract above the pons, very few data are available. We have already seen that anatomists trace the portion of the lateral lemniscus which conveys auditory impulses into the corpus quadrigeminum posterior and into the corpus geniculatum internum.

Weinland¹⁹ has made a collection of 27 cases of lesions of the posterior corpora quadrigemina. In 13 of them deafness was noted, these being all cases in which the lateral lemniscus was involved in the lesion. In some cases the deafness was bilateral, in others it was on the side opposite to the lesion. He is inclined from the study of his own case and of those of Ferrier²⁰ and of Ruel²¹ to believe that a lesion of one corpus quadrigeminum posterior causes deafness in the opposite ear. In his case the right ear became gradually deaf, not from any ear disease, and the tumor had destroyed the left lateral lemniscus. In the case of Ruel the same was true. In the case of Ferrier the tumor destroyed the right corpora quadrigemina posterior and lateral lemniscus and the left ear was deaf.

In the 14 cases of lesion of the posterior corpus quadrigeminum in which no deafness was found, he shows that either the lesion did not reach the lateral lemniscus or there was no record of an examination of hearing.

Oppenheim and Bruns in their works on brain tumors reproduce these statements of Weinland but do not add any observa-

¹⁹ Arch. f. Psych., XXVI, 375.

²⁰ Ferrier, Brain, 1882, April.

²¹ Ruel, Neurol. Centr., 1890, p. 192.

tions of their own and I have failed to find in literature any study of lesions of the corpora quadrigemina posterior except that of Weinland.

A similar lack of observations is encountered when an attempt is made to trace by pathological records the course of the acoustic tract through the corpora geniculata interna and thence to the temporal lobes. The investigations of von Monakow of the connections of the optic thalamus by the experimental methods upon animals prove conclusively that the corpora geniculata interna send their fibers to the cortex of the temporal convolutions. Limited lesions of these bodies or of the tracts proceeding from them are rarely if ever observed in man, hence we must be content with relying exclusively upon the facts afforded by anatomy and physiology for our data. There is every reason, however, to believe that some future observer will establish the fact that partial deafness of the opposite ear may be caused by a lesion of the corpus geniculatum internum or of its radiation outward to the temporal lobe.

It seems somewhat superfluous to state that central deafness, by which I mean deafness due to a lesion of the auditory tract, may develop either suddenly, as the result of a vascular lesion, or gradually, as the result of a tumor. Yet this assertion seems necessary in view of a recent article by Cornet²² who divided sudden deafness into two categories only; first, febrile, due to purulent inflammation in the labyrinth, and, second, afebrile, which he says may be caused either by wax in the ear, by syphilitic lesions of the labyrinth or by hysteria; thus ignoring entirely the possibility of a nerve lesion.

That a gradually advancing deafness of nervous origin and due to lesion in the auditory nerve from atrophy of its ganglionic cells of origin in the labyrinth may occur, especially in connection with tabes, has long been recognized clinically, though pathological proof has only recently been forthcoming, as shown by Bruhl²³ and Hammerschlag.²⁴

In conclusion it is evident that it is possible to divide the pathology of deafness into four definite categories, each having characteristic symptoms of its own, and each being inevitably

²² Archives gen. de Méd., 1906, II, p. 2329.

²³ Zeitschrift für Ohrenheilkunde, LII, 232, 1902.

²⁴ Zeitschrift für Ohrenheilkunde, LVI, 126, 1906.

associated with other symptoms which aid in determining the location of the lesion: (1) Labyrinthine deafness with associated vertigo; (2) acoustic nerve deafness from primary atrophy or associated with tabes; (3) central acoustic tract deafness, associated with symptoms of pontine or crural symptoms; (4) cortical deafness, usually associated with aphasia and presenting the symptom of psychical deafness.