NEGLECT OF EAR-SYMPTOMS IN THE DIAGNOSIS OF DISEASES OF THE NERVOUS SYSTEM

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G. L. WALTON, M.D.

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THE great advance in the knowledge of the nervous system, made during the past twenty years, is due not simply to laboratory investigation, but largely to clinical observation and study.

One branch of clinical work seems, however, to have been left far behind in this connection, for the study of the ear attracts but little more interest to-day among neurologists than it did twenty years ago. This is the more remarkable in that otological research itself, while limited for the most part to specialists, occupies at present a front rank in scientific progress.

That the general practitioner should gain an extended knowledge of the ear is perhaps too much to expect, although it is unfortunate for every-day practice that the interest in this line of work should be at such a very low

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ebb. Dr. Edward H. Clarke,[1] and more recently Dr. Woakes,[2] have, for example, commented upon the lack of knowledge and interest which allows a practitioner to ignore aural symptoms in the exanthemata, and to inform the convalescent from typhoid fever that deafness is a favorable sign; the fact being that a little knowledge of the pathology of the ear, a little forethought, and a moderate degree of experience might have prevented, for example, a hyperæmia from becoming a purulent inflammation, causing such destruction of tissue as to render the ear useless as an organ of special sense, to say nothing of the dangers from extension of the process.

It is not, however, the object of the present paper to discuss the gaps in the general practitioner's knowledge, but to consider briefly the interest which the study of otology should have for the neurologist, and for the practitioner interested in neurology.

It has now become so much a part of the neurologist's education to acquaint himself with the examination of the *eye*, and of his practice to interest himself in it, that the diagnosis of cerebral tumor or locomoter ataxia is rarely made without examination of the fundus oculi, while the mention of "blindness" as a symptom in cerebral disease, unaccompanied by a description of the eyes and the exact nature of the defective vision, would call down severe criticism. Almost every number of the neurological journals contains a discussion either of the course of the fibres of the optic nerve or of some other question regarding the physiology or pathology of the eye.

The result of this enthusiastic study is that as large a part perhaps of neurological advance is due to ophthalmological research as to any one branch of investigation.

[1] *Am. Jour. Med. Sciences*, January, 1858.

[2] "Deafness, Giddiness, and Noises in the Head." Edward Woakes, M.D., London, 1879.

Now, the situation of the optic nerve is such as to render it peculiarly susceptible to alteration from cerebral lesion, while the relation of the auditory nerve to the brain is not such as to warrant the assumption that the study of its functions would throw an equal amount of light upon nervous pathology. It is also true that the optic nerve is generally eligible for direct examination, while the auditory nerve is not. Notwithstanding these facts, none would claim that the neurologist has nothing to gain from the study of the ear, and it is highly probable that systematic examination of the hearing and of the ear in cerebral disease, whether deafness is suspected or not, would, like the examination of the sight and the eye, not only aid much in actual diagnosis, but add greatly to our knowledge of the central nervous system.

As a recent example of the value of adding aural to ophthalmological study in diagnosticating central nervous disease may be cited the subject of hysteria. Thanks in greatest measure to the efforts of Prof. Charcot and his pupils, this disease has been rescued from the uncertainty and contempt of former times, and has been found as worthy of scientific study as any other, and once subjected to systematic analysis it repays us by solving physiological problems with an accuracy not to be attained in laboratory research.

Prior to the studies at the Salpêtrière hysterical blindness was considered a vague symptom unworthy of careful investigation. It has now been known for some time that this, like the other functional anæsthesias, is subject to given laws and offers marked and individual characteristics, so that amblyopia, with concentric retraction of the field of vision and loss of certain colors has become a pathognomonic symptom. In a similar way hysterical deafness, long noticed but not investigated, has now been shown to follow

equally fixed laws, the hearing through the bone disappearing before that through the air, and that for high before that for middle tones.[1] The writer has already found a knowledge of these peculiarities of great practical value, in connection with other symptoms, in establishing the diagnosis of functional anæsthesia,[2] a diagnosis of importance, not only with regard to treatment and prognosis, but particularly in a medico-legal point of view in cases of so-called "railway spine," as pointed out recently by Dr. J. J. Putnam.[3] With regard to the physiological interest of these facts, assuming as we may fairly do that the deafness is due in these cases to disturbance in the cerebral centres, the analogy at once appears between hysterical and senile deafness. In the latter also the hearing through the bone and for high tones disappears first. Previous explanation of these peculiarities, as, for example, by the assumption of impaired bone-conduction in old age, has never been satisfactory; and the same peculiarities occurring in young girls, with no evidence of alteration in the conducting media, but with other evidences of anæsthesia of central origin, tends to show that the hearing for high tones and that through the bone disappear first during impairment of the auditory centres, simply because these are the sounds of which those centres are the least tenacious.

It will not be out of place at this point to observe that the importance of testing the hearing for high tones, as proposed by Dr. Clarence J. Blake,[4] has indeed hardly at-

[1] Deafness in hysterical hemianæsthesia. *Brain*, No. xx, 1883; also *Verhandlungen der physiologischen Gesellschaft zu Berlin*, Feb. 9, 1883.

[2] In one case, published in the *Archives of Medicine*, Aug., 1883, the hearing was examined with König's rods at intervals during convalescence, and it was found that the scale of notes audible on the affected side gradually rose until the rod of 30,000 vibrations could be heard on this side, to that of 35,000 on the other. The diagnosis in this case was settled by the disappearance of the deafness on application of an electro-magnet.

[3] *Boston Med. and Surg. Journal*, Sept. 6, 1883.

[4] Summary of the results of experiments on the perception of high musical tones. Trans. Am. Otological Soc., 1872. Diagnostic value of high musical tones. Trans. Am. Otological Soc., 1873.

tracted sufficient attention, even among otologists, though already, in 1877, the "méthode de Blake" was described by Guerder [1] as furnishing the *key* to diagnosis between deafness resulting from lesion of the nervous apparatus and that resulting from impaired transmission. Dr. Blake has shown, by testing for high tones after incision of the membrana tympani, that the inner ear is capable of conveying, and the brain of perceiving, normally, at least 80,000 vibrations, although the ordinary limit while the membrane is intact may be placed at 40,000 vibrations. It is certainly remarkable that the high tones have received so little attention, and that the König's rods are so little in general use for diagnosis.

The attention of the writer was particularly drawn some time ago to the lack of attention given the ear in diagnosticating cerebral disease, while looking through the neurological journals. Attention having been attracted to the subject, the search for cases of cerebral disease has been continued at some length through leading German, French, English, and American journals, simply to observe how often note is taken of the aural symptoms, and when taken, how often systematic examination of the ear is made.

The result has not been satisfactory. The ocular symptoms are rarely neglected; the condition of the fundus is almost invariably reported where it can throw the least light on the nature of the lesion. It is needless to say that the statement is rarely found that defective vision exists, without such information as tends to show whether the difficulty is due, for instance, to opacity of the lens, optic neuritis, or refractive irregularity.

The contrast between this desirable accuracy and the loose statements with regard to the aural symptoms, not so noticeable in the single reports, becomes absurd when a large number are passed in review.

[1] *Annales des maladies de l'oreille, du larynx, et des organes connexes.*

The cases in which ear-symptoms are neglected in the diagnosis of diseases of the nervous system may be classed as follows: (1) The cases in which no note whatever is taken of the condition of the hearing, although the presence or absence of deafness would prove of diagnostic value; (2) the cases in which deafness is mentioned as a symptom in disease of the nervous system, without particulars as to its nature and degree, and without sufficient examination of the ear to eliminate defective transmission of sound.

(1) With regard to the first class of cases, those in which the hearing is totally neglected, this is by far the most numerous, and comprises perhaps the large majority of cases of cerebral lesion, such as tumor, hemorrhage, and abscess, to say nothing of hysteria and allied disturbances. Why the auditory nerve in particular should be left out in the otherwise systematic analysis of such cases is not clear, and is only to be explained by the fact that attention has never been especially called to its importance. The fact that the examination of the hearing and of the ears is difficult, is certainly no reason for leaving them altogether out of consideration in reporting cases where they would be of value.

The presence or absence of deafness is, for example, of the utmost importance in diagnosticating lesion of the pons or medulla oblongata, or of the cerebellum. As an illustration of this fact, Seymour has recently reported a case of cerebellar tumor pressing on the pons,[1] in which absolute deafness (unilateral), both for sounds conveyed by air and for those conveyed by the skull, added greatly to the certainty of the diagnosis, which was afterward corroborated by post-mortem examination.

Although this case unfortunately comes under the second class just mentioned, in that no examination of the ear

[1] *Boston Med. and Surg. Journal*, August 30, 1883.

was made, the probabilities were greatly in favor of the tumor as the origin of the deafness, and the case is quoted as a step in the right direction, chiefly because Nothnagel, in his standard work on the diagnosis of cerebral disease (edition of 1879), has stated that no authentic case is on record of deafness resulting from cerebellar disease.[1] The same author, in considering lesions of the pons, remarks,[2] that considering the situation of the auditory nerve it is curious that deafness is rarely found as a symptom. The fact of its non-mention in a long series of cases he regards as proof of its rarity, for, he says, it is hardly probable that a marked degree of deafness could go unnoticed. This by no means follows, for deafness is a symptom which, in the majority of cases, unless carefully sought for, escapes the notice, not only of the patient himself, but of the medical practitioner, many persons going through life unaware of the fact that they are almost totally deaf in one ear. This point will be alluded to again farther on. By far the most probable explanation of the seeming rarity of deafness as a symptom in tumors and hemorrhages in the pons and cerebellum is, that the hearing is rarely examined in such cases.

(2) Examination of the second class of cases (those in which deafness is noticed but not accompanied by description of its degree and nature, or by examination of the ear) reveals the lack of interest—not to say lack of knowledge—prevalent in this branch of symptomatology.

Omitting the cases reported by otologists, a case is rarely found in which a systematic examination of the ear and of the hearing is made, so that in many cases the reader is even left in doubt as to whether the deafness is really due to the cerebral lesion, or, perhaps, to a plug of cerumen, or catarrh

[1] "Topische Diagnostik der Gehirnkrankheiten," von Dr. Hermann Nothnagel, Berlin, 1879, S. 595.

[2] *Ibid.*, S. 154.

of the middle ear. This neglect is the more noticeable in that the writers include observers otherwise most accurate, for it seems to be only the most careful clinicians who test the hearing at all. By these, the symptom deafness is repeatedly mentioned in connection with the various disturbances of the central nervous system—such as locomotor ataxia, trauma, new growth, and hysteria,—without the least record of the hearing for different tones, or for that through the bone, and often with either no examination of the ears or a very superficial one. In some exceptionally careful reports appears the statement, "membranes normal," as if this fact absolutely established the central origin of the deafness, whereas it is well known to otologists that an apparently normal membrana tympani may have beyond it a middle ear so far injured by catarrhal inflammation as to cause marked loss of hearing.[1] If, in such a case, the hearing through the bone and that for different tones is unimpaired, a central origin for the deafness is eliminated, and even if the hearing through the bone is lost, the fact is not yet established that the nervous auditory mechanism is affected, for catarrh of the middle ear, though generally apparently increasing the intensity of sounds conveyed by the bone, by preventing the passage outward of the vibrations, sometimes suspends the hearing for sounds conveyed in this way (probably by causing anchylosis of the stapes, thus preventing the vibration of the labyrinthine fluid).

Even when disease of the outer and middle ear has been eliminated, we are by no means justified in jumping at once to the cerebral auditory centres, or even to the auditory fibres in the brain, for there still remain the intricacies of the labyrinth, and the course of the auditory nerve to the brain, either of which may be the seat of lesions causing

[1] Urbantschitsch: "Traite des maladies de l' oreille," trad. franc., 1881, p. 267.

loss of auditory function. In consideration of these facts how little scientific is the report, for example, of a case of locomotor ataxia with the symptom "deafness" mentioned without further particulars, and how unfortunate that the diagnosis of probable traumatic lesion of Ferrier's auditory centre should be made, as in a case recently reported in the journal *Brain*, with no examination of the ear until some months after recovery (which in the case alluded to, by the way, revealed old purulent inflammation of the middle ear), and no more careful test for the hearing at any time than that for the watch by ear and through the skull, a test so untrustworthy as to be discarded by many otologists.

Amongst other inaccuracies may be noticed the fact that the patient's own statement is evidently often relied on in eliminating the question of deafness as a symptom. Now, the patient's opinion on this point, even when he has noticed it, is generally unreliable, a deafness from catarrh or cerumen of several years' standing being often considered by the patient to have commenced a few months previously. This illustrates the well-known fact, that a considerable degree of deafness may go unnoticed for an indefinite time; and, indeed, provided it be unilateral, the deafness may become total, and yet remain unperceived, until suddenly revealed by accident, as by rolling over upon the well ear in bed.

These facts render the custom of questioning the patient with regard to his hearing of very little value as compared with objective tests.

Even the tests for hearing by the voice, tuning-fork, and rods, are subject to such inaccuracies as to require careful study, considerable experience, and repetition in the given case. For example, the patient generally understands so well what is said to him by gesture, look, and the motion of the lips, as to render the voice test unreliable unless these

sources of error are removed. Deaf children are often brought to the aural clinics who not only deceive their parents, who are generally willing to be deceived, but who would deceive the medical practitioner unless he exercised great care, because they turn so quickly when a noise is made that it seems as if they must have heard it. The same children will, perhaps, take absolutely no notice of the shrillest sound if made stealthily behind the head by an experimenter who remembers that the field of vision extends laterally over not far from 180 degrees when the head remains quiet, and much farther when it is continually in motion.

These single instances of the many sources of error familiar to otologists, will serve to remind us of the need of careful study before making scientific reports of the hearing power in cerebral disease.

As an example of lack of care among neurologists in eliminating trouble in the ear itself, Lucae,[1] as early as 1866, exhibited the peripheral auditory apparatus of two patients who had died from locomotor ataxia, demonstrating the real seat of the deafness, and calling attention to the fact that deafness in this disease should not be attributed to central disturbance without careful examination of the ears. Since that time, however, deafness has been in many cases reported in connection with locomotor ataxia, sometimes with no examination, and sometimes with but the most casual examination of the ears, and Pierret[2] has even suggested, on the most insufficient evidence, that degeneration of the auditory nerve is one of the early symptoms of this disease, the fact being probably that if peripheral disturbance were carefully eliminated, according to Lucae's suggestion, the cases of deafness from locomotor

[1] *Verhandl. der Berl. med. Gesellsch.*, Bd. i, p. 127, 1866.

[2] *Revue mensuelle*, fevrier, 1877.

ataxia would be reduced to a minimum. Careful search by the writer through a series of forty cases has, indeed, failed to reveal a single case in which central deafness could be established, though the majority of the patients were more or less deaf.

As another example, may be cited the frequency of the diagnosis and treatment of "Menière's disease" without examination of the ear, while, in fact, the so-called Menière's complex of symptoms (deafness, noises in the ear, nausea, and giddiness, with or without tendency to fall in a particular direction) occurs so frequently in connection with disease of the external and middle ear, that the cases are comparatively rare in which otologists refer these troubles to lesion of the auditory nerve or its terminations.

It is not the object of this paper to contend that the skill of an otologist should be added to the already varied requirements for neurological training. It is rather to offer the suggestion that the ear deserves an interest at least approximating that accorded to the eye in the diagnosis of nervous diseases. It is certainly not too much to expect that every practitioner, whether neurologist or not, should either refer cases to a specialist or practise the examination of the ear and hearing to such an extent as to avail himself at least of the aid gained from the appearance of the membranes, the patency of the Eustachian tubes, and the hearing by air and bone by the various tests, as well as the hearing for different tones, before making a diagnosis of lesion of the nervous auditory apparatus. And further, it is to be hoped that the time is not far distant when reports of cerebral disease ignoring the condition of the hearing and the examination of the ears, will be considered as incomplete as they are at present without record of the condition of the eyes.

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