THE SYMPTOMATOLOGY OF ACOUSTIC TUMORS
WITH SPECIAL REFERENCE TO
ATYPICAL FEATURES

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The symptomatology of tumors of the cerebellopontine angle, particularly the acoustic neuromas, has been worked out carefully over a period of many years, beginning with the first case history described by Lévêque-Lasource in 1810. Although this may have been a meningioma, the symptoms were well tabulated. The earliest complete story was that given by Cruveilhier in 1835, in his description of a patient who was proved at autopsy to have a tumor of the eighth cranial nerve. Apparently the first diagnosis of an acoustic neuroma in a living patient was made by Stevens in the United States in 1879. From then until the appearance of the monograph by Cushing in 1917, the reports concerning the clinical syndrome became increasingly frequent.

As summarized by Cushing, the syndrome consisted of a symptomatic progress in the following order:

1. Auditory and labyrinthine disturbances.
2. Occipitofrontal pains and occipital discomfort.
3. Incoordination and instability of cerebellar origin.
4. Evidence of adjacent cranial nerve involvement.
5. Indications of increasing intracranial pressure.
6. Dysarthria and dysphagia.
7. Cerebellar crises and death.

In the majority of cases these symptoms came in this definite chronologic order and the syndrome was verified so often that it was thought to be almost infallible. In fact, we too believe that if the complex is not typical some other type of tumor is likely to be present.

The surgical treatment of these growths has changed radically in the last twenty years, so that the unilateral approach and total removal have become accepted more and more as the best mode of attack. This method was described first by Dandy in 1924 and enlarged upon by him in 1934. Accordingly, it becomes obvious that an accurate diagnosis is more than ever essential, especially since a decompressive operation for a nonremovable growth under unilateral exposure has been unsatisfactory. For this reason we examined the histories of the patients seen at the Lahey Clinic from the beginning of the neurosurgical service, November 1, 1932, to January 17, 1942 to determine the reliability of the various symptoms and signs, as well as to gain some idea as to how frequently special methods of examination such as lumbar puncture, special roentgenologic examination and air
studies are necessitated in patients suspected of harboring these tumors.

Our series of verified acoustic neuromas to January 17, 1942 totals 54 patients, but only 42 of these came to the Clinic for their primary operation. Those who had been subjected to previous surgery elsewhere were excluded from this study. The patient's symptoms and signs were considered from the standpoint of subjective and objective manifestations. Obviously, the complete chronologic story rarely is volunteered by the patient and can be obtained only by careful questioning. The chief findings will be discussed as to their frequency, together with their duration before the patient was admitted to the hospital.

SUBJECTIVE DISTURBANCES

Auditory and Labyrinthine. Deafness was present in 39 patients (92.8 per cent), absent in 2, and unknown in 1 because of coma. It was bilateral in 1 patient. The duration of deafness varied from six months to ten years (except in 1 patient in whom it had been present for thirty-two years, probably from other causes). Excluding the thirty-two year case, the average period of deafness was two years and ten months.

With the exception of the 2 cases in which deafness was absent and 4 cases in which it followed tinnitus, deafness was preceded by other symptoms in but 1 patients. In 1 of these, trigeminal disturbances occurred earlier, and in the other, unsteadiness in walking. These 2 exceptions, however, should be borne in mind since they are a definite departure from the almost universal chronology.

Tinnitus was present in 26 patients and absent in 16. It was present on the side of the lesion in 24 instances, bilateral in 1, and on the opposite side in 1. Its duration was from two weeks to over six years. So far as can be judged from the records, tinnitus preceded deafness in only the 4 patients previously mentioned, these two symptoms usually becoming manifest simultaneously. In the 2 patients without deafness, unilateral tinnitus had developed a few weeks prior to their admission.

Perhaps the most striking point about the symptom of tinnitus is that it was absent in 16 patients (38 per cent), whereas it has always been regarded as one of the most frequent and early manifestations of acoustic tumors.

Instability. Thirty-one patients (74 per cent) were aware of staggering or unsteady gait. In 10 cases it was absent, and in 1 case questionable. Only occasionally was it toward the side of the lesion. Instability was one of the earlier symptoms, having been present from two months to three years, with an average for all patients of one year and three months.

Headache. As would be expected, this was a frequent complaint, being present in 34 patients (81 per cent). Its duration was from ten days to ten years, with an average of one year and three and a half months. Excluding the 1 patient in whom headache had been present for ten years, the average was just over one year. In 4 patients headache was said to have antedated all other symptoms, but whether it was due to the tumor is questionable.
Trigeminal Symptoms. Hypesthesia or paresthesia of the face which, with one exception, was on the side of the lesion, was noted by 27 patients, and had been present from five months to five years.

Concerning a possible relationship between headache and pressure upon the trigeminal nerve, it is perhaps noteworthy that all patients without trigeminal symptoms had headache for only a short time or not at all; also, that patients without headache had trigeminal symptoms for six months or less, with one exception in which the duration was three years.

The significance of this observation is not at once apparent. However, it may be inferred that the headache of a patient with an acoustic neuroma is perhaps less significant of increased intracranial pressure than of encroachment on the sensory root of the fifth cranial nerve. This becomes even more possible when analysis shows that in 81 per cent of our cases the duration of the two complaints was relatively similar.

Visual Impairment. Twenty-five patients (59.5 per cent) complained of blurred or double vision. In 14 of these, the complaint had been present for a shorter time than had the headache, and in 10 patients it appeared after the trigeminal symptoms. In 21 cases, the visual symptoms were associated with obvious choked disks or secondary atrophy and blindness. Analysis of the figures fails to show visual symptoms to be any more reliable criteria of increased intracranial pressure than headache, since 13 patients without visual complaints had choked disks and 6 without headache likewise showed papilledema. Four patients with both headache and visual symptoms had no elevation of the optic disks.

Vertigo. This, either constant or in attacks, was conspicuous by its infrequency, being noted in only 13 patients (31 per cent). Its duration was from two weeks to two years.

Subjective complaints related to adjacent cranial nerve involvement, except the fifth, were almost absent. In rare instances dysarthria, dysphagia, squint or facial twitching had been noted by the patient.

OBJECTIVE MANIFESTATIONS

Nystagmus. From the objective standpoint, the only finding uniformly present was nystagmus. This varied in intensity with different patients but could be elicited in all. It was horizontal in every patient and often vertical as well, but whether the excursions differed in rapidity was not noted in all patients. In the 33 cases in which nystagmus was described, there were slower or coarser excursions on deviation of the eyes toward the side of the lesion.

Labyrinthine Impairment. In 9 patients caloric examination of the external auditory canals was not performed, but in the 33 who had this examination there was an absence of response from the labyrinth on the side of the lesion, in 1 patient the loss being bilateral. Of the 2 patients with normal hearing, 1 showed a dead labyrinth on the side of the tumor, and in the other patient the caloric test was not done.
Impairment of Hearing.* Deafness was present in 39 patients, and could not be determined in 1 because of coma. In 2 of the 39 it was very slight. Two patients had normal hearing bilaterally. The condition was present only on the side of the lesion, except in 1 patient in whom it was bilateral because of chronic otitis media.

Papilledema. In 32 cases the optic disks showed obvious choking, or choking combined with secondary atrophy. The fundi were normal in 8 patients, and in 2 there was questionable edema. In 4 patients pressure on the optic nerves had been allowed to continue for such a long period before they were referred for surgery that they were either blind or had scarcely useful vision at the time of their admission to the hospital.

Trigeminal Involvement. Thirty-three patients had evidence of impaired trigeminal function either as a demonstrable hypesthesia or an absent corneal reflex on the side of the lesion. Seven patients showed no trace of trigeminal involvement, and in 2 it was questionable.

Other Cranial Nerve Involvement. The infrequency of true ocular muscle palsy in cases of acoustic neuroma was noted by Horrax and Buckley, and in only 4 of our patients was there a definite sixth nerve weakness. Pupillary changes or mild difficulty in conjugate movements, especially upward, were present in 11 other patients. A definite peripheral facial weakness was noted in 12 cases, and a mild or questionable loss was present in 15. Eight patients had evidence of ninth and twelfth nerve involvement, but this was severe in only 2 and questionable in 2 others. Accordingly, it becomes apparent that involvement of the neighboring cranial nerves, except for the fifth, is less common than would be expected, especially when one considers their number and proximity.

Evidences of Incoordination. Unsteadiness in walking was objectively present in 32 patients and absent in 9. Romberg’s sign was positive in 26 cases and absent in 15. It was impossible to test 1 patient on account of coma.

Finger-to-nose ataxia was manifested in 38 patients and was not elicited in 4. It was designated as greater on the side of the lesion in 31 patients.

Suboccipital Tenderness. Suboccipital pain or tenderness to pressure or some form of uncomfortable feeling in the back of the head or upper portion of the neck was a fairly common finding, being present in 24 of the 42 patients. It was not of diagnostic value as to the side of the lesion.

VENTRICULOGRAMS

In 8 cases a ventriculogram was considered necessary to determine definitely the need of a suboccipital approach or to differentiate tumor from some other condition. In only 1 patient did the air study lead us to suspect an acoustic neuroma by deformity of the occipital horn of the lateral ventricle, as described by Stone and Schulze. A summary of this case follows:

* In many of the patients hearing tests were made with the audiometer, but in all cases careful tests by the usual clinical methods including air and bone conduction by the tuning fork were carried out, usually by several observers. In the two patients in whom hearing was said to be normal this was so recorded by the audiometer in one patient and in the other the light rubbing of fingers and the faint ticking of a watch could be heard equally well with either ear when the opposite ear was tightly plugged.
Case 1. M. B., a 51-year-old white woman, was admitted to the New England Deaconess Hospital on September 21, 1940, with a complaint of headaches since May 1940. She had had two miscarriages, seasonal hay fever and two minor head injuries in 1939. The headaches were at first occipital in origin with radiation forward, but finally became bitemporal and frontal. They came on suddenly, lasted a few minutes and disappeared as suddenly. During the more severe attacks the patient noticed some blurring of the eyesight. In the summer she began having attacks in which she would suddenly fall to the floor, lose consciousness and jerk her arms a few times. She had no other complaints, but stated that she had been somewhat deaf in the left ear for two years.

General physical examination revealed obesity, normal blood pressure, and negative heart, lungs and abdomen. Pelvic examination showed a large fibroid uterus. There was no scalp tenderness or neck rigidity. Laboratory studies showed a normal blood count and a negative Hinton reaction. There was a faint trace of sugar in the urine on two examinations.

Neurologic examination demonstrated bilateral choked disks and normal pupils. There was nystagmus to the right and left, which was slower on left lateral gaze. There was no facial weakness or hypesthesia. However, there was a slight left-sided deafness. Regrettably, caloric studies were not done. The deep reflexes were a little more active on the left. There was no ataxia, but on trying finger-to-thumb approximation, she was a little slower on the left than on the right. A moderate degree of generalized hypotonia was also noted.

It was felt that she was suffering from an intracranial tumor, probably midline cerebellar in location, but the signs were too few to warrant surgery without an air study. A lumbar puncture was not done because of papilledema.

On September 25, a ventriculogram was made and this showed uniformly dilated lateral and third ventricles, but there was a slight elevation of the left occipital horn. The third ventricle was also shifted to the right and slightly superiorly at its posterior end. These changes were thought to be sufficient to warrant a left suboccipital exploration for an acoustic neuroma. This was performed the same day and what was described as a moderate-sized tumor was removed completely without difficulty.

The postoperative course was amazingly uneventful and she was discharged on October 16, moderately ataxic on the left side but able to be up and about. She had the usual complete facial paralysis but refused an anastomosis at that time. On May 5, 1941, she returned and a hypoglossal-facial anastomosis was performed. There was at this time a moderate-sized collection of fluid under the scalp flap which was aspirated. She stated that this became somewhat tense at times but that she felt well and was doing her housework.

Comment. In retrospect it is obvious that not sufficient attention was placed upon the slight but definite deafness of the left ear, probably because there was no accompanying trigeminal hypesthesia. Caloric examinations doubtless were omitted because the deafness was so slight, but in all certainty they would have shown a dead labyrinth on the left. This, of course, would have clinched the diagnosis. It is probable too that if a lumbar puncture had been performed, and this had disclosed an elevated protein content of the spinal fluid, the patient might have been spared the ventricular study. However, the usefulness of this procedure in occasional somewhat obscure cases must always be considered.

In a second case in which ventriculography was utilized, evidence of a posterior fossa tumor was definite from air study, but an acoustic neuroma was not expected. The following case is representative:

Case 2. V. S., a 37-year-old white woman, was admitted to the New England Deaconess Hospital on April 5, 1937, with the complaint of headaches for four years and dizziness and double vision for one year. The family and marital history were negative. She had had a toxemia of pregnancy many years before and a minor head injury three years previously.
The headaches were of two kinds. The first type came on when she lifted her head while lying down and was a dull ache of short duration, appearing in the right suboccipital region. The other type came on about once a month and was a sharp prostrating pain appearing like two prongs, starting in the back of the head and radiating forward on each side.

One year before admission, she noticed staggering when she walked, with a tendency to deviate to the left, and some clumsiness of the left hand. Two months later she developed double vision and four months later a dull ringing noise in the right ear. Four months preceding admission she noticed some flashes of pain in the right cheek and recently some tingling on both sides of the mouth.

General physical examination showed an apprehensive patient with no abnormalities noted in the mouth, throat, lungs, abdomen or genito-urinary tract. Routine blood and urine examinations were normal.

Neurologic examination revealed chronic, bilateral choked disks but normal visual acuity. There was nystagmus to the right and left, with slower excursions on left lateral gaze. There was slight hypesthesia of the left side of the face but no facial weakness and no loss of hearing. Caloric examination was not made. The lower groups of nerves were normal in activity on both sides. There was ataxia of the left arm and leg on doing the various tests. There was a positive Romberg's sign, and the gait was ataxic with deviation usually to the left. Deep reflexes were overactive throughout but were equal.

The impression was that this patient was suffering from a midline cerebellar or a posterior third ventricle tumor. On April 8, 1937, a ventriculogram showed uniformly dilated lateral and third ventricles. On bilateral suboccipital exploration the same day, a left acoustic neuroma was exposed and removed completely. It was felt that the facial nerve was preserved during the removal and this has subsequently proved to be correct. The growth itself was of only moderate size, but there was a large arachnoid cyst overlying it.

After an uneventful postoperative course, she was discharged on May 1 in good condition and with a little motion beginning to appear in the left side of the face.

Comment. This case again represents the importance which would doubtless have been placed upon an absent labyrinthine response if a caloric examination had been performed. It was probably omitted because the patient had no deafness, a fact which in itself militated against the diagnosis of an acoustic tumor. However, it is the occasional departure from the usual that needs emphasis, and obviously when patients, like this one, have even a slight trigeminal involvement along with cerebellar symptoms, tests of labyrinthine function should certainly be a part of the examination.

ROENTGENOGRAMS

There was no case in which roentgenologic examination of the petrous ridges or of the internal auditory meati was the final diagnostic criterion, but the roentgenograms showed positive findings in 9 of the 15 cases in which they were made. The examinations used were first described by Carr in 19171 and Towne in 1926.11

LUMBAR PUNCTURE

In our opinion, "routine" lumbar punctures in patients harboring intracranial tumors are not only contraindicated but are definitely dangerous, particularly so when there is a lesion in the posterior fossa because of greater herniation of the cerebellar tonsils through the foramen magnum. However, in certain instances the spinal fluid findings may be of distinct diagnostic aid, and if the procedure is carried out in a hospital where imme-
diate operative intervention can be instituted if necessary, the danger is at least greatly minimized.

In two of our patients lumbar puncture contributed distinctly to the correct diagnosis. One of these cases is summarized here. In a third patient, a spinal puncture done elsewhere precipitated a train of symptoms which a few days later was diagnostic of an acoustic neuroma. Merritt has stated that acoustic neuromas, of all the brain tumors, are most prone to produce elevation of the protein content of the fluid obtained by lumbar puncture. Nine of our patients had the procedure performed here or elsewhere, and all showed proteins of 100 mg. per cent or more. In the following case the protein elevation gave us the final clue between an acoustic neurona and an intrapontine lesion.

Case 3. L. S., a 63-year-old white woman, was admitted to the New England Deaconess Hospital on March 18, 1940, with a complaint of numbness in the left side of the face and decreased hearing in the left ear for one year. At the onset of the trouble she noticed some noises in the left ear, but these stopped after a time. The deafness and numbness had slowly progressed. At the time of menopause, fifteen years before, she had a great deal of nervousness and this had recurred and become very severe in the month before she was admitted. She had had sick headaches when younger but none for thirteen years. In the last three years she had had frontal headaches and a feeling of fullness around the left eye which went away when she was busy with housework. She had had teeth extracted for the facial trouble, and a fairly severe anemia developed which was relieved by treatment. The remainder of the history was not significant.

General physical examination showed dehydration, evidence of weight loss and peripheral arteriosclerosis. The blood pressure was 148 mm. of mercury systolic and 90 mm. of mercury diastolic. Laboratory studies revealed a trace of sugar in the urine, the red blood cell count was 4,910,000, the white count 8,950, and the hemoglobin 101 per cent. The blood Hinton was negative. A roentgenogram of the skull revealed calcification in both internal carotid arteries.

Neurologic examination showed mild mental confusion, memory loss, poor insight and orientation. The optic disks were blurred nasally and the pupils were unequal, the left being larger than the right, but both reacted well. Nystagmus, horizontal in direction, was present both on looking to the right and left, but was slower and of wider excursion on left lateral gaze. There was definite hypesthesia in all three divisions of the left fifth nerve, most pronounced in the third. A slight left peripheral facial weakness was present as well as a mild right central one. The left ear was deaf, and the caloric response was absent on that side. The other cranial nerves were negative, and there was no localized weakness of the extremities or sensory change. Deep reflexes were active throughout but a little more so on the right and the superficial reflexes were not significant. Coordination tests revealed a little ataxia on the left, an unsteady gait and a tendency to fall backward in the Romberg position.

It was our impression that the patient was suffering from an intrapontine or left cerebellopontine angle tumor in addition to cerebral arteriosclerosis. Lumbar puncture revealed a pressure of 100 mm. of water and xanthochromic fluid containing 480 mg. of total protein per 100 cc. Reexamination of the roentgenograms showed erosion of the medial end of the left petrous ridge.

With these findings a diagnosis of left acoustic neuroma was made. On March 27, 1940, a left unilateral suboccipital craniotomy was performed and a moderate-sized acoustic tumor was removed completely without difficulty. The internal auditory meatus was curetted and the facial nerve sacrificed.

The postoperative recovery was slow, chiefly because her mental status remained unimproved and she refused to eat. She also developed cystitis due to total incontinence. She was discharged in fair condition on May 17. The left eyelid was closed permanently by suture on September 18, 1940, in order to take care of a severe keratitis and at this time her mental status was still only slightly improved.
SUMMARY

Although the specific symptomatology and particularly the chronologic development of symptoms is highly important in the diagnosis of acoustic tumors, nevertheless the occasional absence of certain usual features, especially deafness, should be emphasized.

The subjective symptoms and objective findings in a series of 42 patients having verified acoustic neuromas have been analyzed. The usual chronology of the patient's complaints in this series is in essential agreement with the summary given by Cushing in his monograph in 1917. Certain features, however, bear emphasis. Tinnitus, for instance, was absent in 38 per cent of patients, and although it was noticed usually at about the same time as impairment of hearing, it preceded deafness in only 4 instances. The close association of trigeminal nerve involvement with headache was of interest, and it may be that headache in a patient with an acoustic tumor is less significant of increased intracranial pressure than of pressure upon the fifth cranial nerve. This is further borne out by the fact that 4 patients who had both headache and visual symptoms showed no papilledema.

One significant objective observation was the presence of nystagmus in every patient. It was slower toward the side of the lesion in all instances in which it was recorded.

Caloric examinations were omitted in 9 patients, but an absent response from the labyrinth on the side of the lesion was ascertained in every case in which the test was carried out. The value of an absent response from one side as diagnostic of an acoustic growth in patients having cerebellar signs and symptoms hardly can be overemphasized. Caloric tests should always be performed whenever there is the slightest suspicion of an acoustic neuroma, and not infrequently may obviate the necessity of a ventriculogram in doubtful cases. The latter, however, may still be necessary in rare instances, and the same is true of a diagnostic lumbar puncture.

REFERENCES

2. Cruveilhier, J. Cited by Cushing6 (p. 4).
7. Lévéque-Lasource, A. Cited by Cushing6 (p. 3).