Neoplasms of the Internal Auditory Canal*

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Introduction

Histopathological studies of tumors of the temporal bone are scarce. The authors examined a relatively large number of sectioned human temporal bones in search of small asymptomatic acoustic neurilemmomas and have reported the findings (Leonard and Talbot, 1970). During that search, several other neoplasms were encountered in the internal auditory canal. This report presents a clinical and pathological review of these lesions.

Method

Serially sectioned temporal bones from 490 autopsied patients were studied. Eight-hundred eighty-three temporal bones were considered suitable for inclusion in the study, specifically to assess the internal auditory canal for the presence of any type of neoplasm. The difference of almost 100 bones (883 vs 980) is accounted for in one of the following ways: One of the pair of bones was missing (30 bones); tissues were absent from the internal auditory canal on one side of a pair (49 bones); and the tissues were absent from the internal auditory canal on both sides of nine autopsy specimens (18 bones). The autopsy specimens were collected from The Johns Hopkins Hospital and the Baltimore City Hospital during the years 1929 to 1941—the majority of the bones gathered between 1929 and 1934. Approximately three-fourths of the specimens came from The Johns Hopkins Hospital. The specimens came from an unselected hospital population, but many of the patients were from the neurosurgical service. All of the bones were processed in The Johns Hopkins Otological Research Laboratory by methods standardized by the late Stacy Guild. Most of the bones were sectioned in a vertical direction. All patients had audiograms (pure tone, air conduction, and bone conduction) performed while alive. All bones collected and processed during the years named were studied.

The study consisted of microscopic examination of every mounted section of the temporal bone in which any part of the internal auditory canal was included.

Findings

Tumors encountered included four small asymptomatic neurilemmomas, three known neurilemmomas that were operated upon and died postoperatively, two meningiomas, one medulloblastoma, one eosinophilic granuloma, and one astrocytoma.

Asymptomatic Acoustic Neurilemmoma

Four of these small lesions were found. These are described in detail in Leonard and Talbot (1970). Related findings are included in the other report.

Postoperative Acoustic Neurilemmoma

Case One (Pathology Number 12411). This 42-year-old male had noted the gradual onset of hearing loss in the left ear four years prior to treatment. This had progressed, along with tinnitus, until the loss became complete. About one year prior to treatment he noted frontal headaches, blurred vision, vertigo, and difficulty in walking. In the few months before coming to the hospital he noted blindness in the left eye, numbness in the left half of the face, partial loss of taste and smell, and falling to the left.

Physical examination revealed weakness in the left leg, positive Romberg to the left, total blindness in the left eye, loss of taste and smell (left), no caloric response on the left, and a 50 dB flat sensorineural hearing loss on the left. Diagnosis was a left cerebellopontine angle tumor, and partial removal was accomplished via an occipital craniotomy. The patient expired a few hours postoperatively.

The tumor was an acoustic neurilemmoma. The temporal bones (Fig 1, A and B) showed that the tumor almost filled the internal auditory canal and had spread all the way to the spiral ligament. There was little bone destruction.

Case Two (Pathology Number 12657). This 40-year-old female was admitted to the hospital because of headaches and left sided deafness. She had noted left tinnitus about three years previously, onset of hearing loss about a year later, and total deafness on
Fig 1 (A)—Path. No. 12411. Neurilemmoma completely filling the internal auditory canal (IAC). Facial nerve is beginning to exit from the canal at lower right. ×11. H & E stain.

Fig 1 (B)—Path. No. 12411. Slide 1(A), at spiral ligament, shown at magnification of 165×.

the left one year prior to admission. Six months previous to admission she developed severe occipital headaches, momentary bouts of blindness, left facial numbness, and staggering to the left on walking. She had one month of occasional vomiting, and one week of difficulty swallowing.

Physical examination showed a positive Romberg to the left, ataxia, diplopia, left facial anesthesia, no spontaneous nystagmus, a greatly decreased left caloric exam, and a sloping sensorineural hearing loss on the left with approximately 60 dB hearing loss. Masking was not used; we believe this represents a nonresponsive cochlea with a shadow curve from the opposite side.

Diagnosis was a left cerebellopontine angle tumor. Partial removal was accomplished via an occipital craniotomy and the patient expired of surgical complications. The tumor was an acoustic neurilemmoma. The temporal bones revealed the tumor mass occupying about one-half of the internal auditory canal. The tumor had not caused much bony destruction and the facial nerve was intact.

Case Three (Pathology Number 12669). This 60-year-old male was admitted because of recurrent right facial pain. About two years prior to admission, he began having episodes of pain that extended from the right supraorbital area to the occiput. Pain became almost constant; six months prior to admission he was operated on for "tic douloureux" and was relieved for about two months. Twenty years earlier, he had had a right myringotomy for the removal of coal dust. A few months after this procedure, he lost all hearing on the right side.

Physical examination revealed decreased smell, slight diminution of sensation in the right trigeminal nerve, diminished right corneal reflex, no hearing on the right, and normal rotational vestibular tests. Diagnosis was a right cerebellopontine angle tumor, and at operation a large, orange sized tumor was removed. The tumor, a neurilemmoma, indented the brain stem. Removal was difficult and the patient expired a few hours later. The right temporal bone revealed residual tumor along the periphery of the internal auditory canal, extending to Scarpa's ganglion. The facial nerve was intact.

Meningioma

Case One (Pathology Number 16014). This 76-year-old housewife died a few hours after admission; cause of death was acute gastroenteritis. Past medical history revealed that the patient had had seven miscarriages, and seven of her children had died in early infancy due to various causes. At age 66, she was found to have central nervous system lues. History further revealed that she had had, for many years, unsteadiness while walking and particular difficulty while walking in the dark. In the four years prior to
her death, she developed tinnitus and a progressive hearing loss in the left ear to the point that she could hear only loud shouts. In the three months prior to her death, there had been occasional drainage from the left ear.

Physical examination revealed a central perforation in the anterior inferior quadrant of the left tympanic membrane, and there was active suppuration in the middle ear space. Audiogram consisted of only a pure tone, air conduction study. There was a flat 20 dB loss on the right, and a sloping 65 dB loss on the left, when measured with adequate masking. Neurological examination was sketchy, but revealed that the gait was staggering, and the Romberg test was markedly positive. Because of the acute illness at the time of admission consisting of profuse diarrhea, high fever, and dehydration, no other physical findings could be elicited.

At postmortem examination a complete intracranial autopsy was performed, and temporal bones were obtained. The left temporal bone showed evidence of chronic tympanomastoiditis, and there was a large, central perforation in the tympanic membrane. Examination of the left internal auditory canal revealed a small meningioma (4 x 4 mm in diameter) within and confined to the internal canal (Fig 2, A and B). The histological pattern of the meningioma was of the endotheliomatous subgroup, and it contained numerous vascular channels and psammoma bodies. We believe this tumor had its origin from an arachnoid villus within the left internal auditory canal. The tumor did not compress the VIIth or VIIIth nerves, nor did it invade them. This case is discussed in somewhat greater detail in a report by Nager (1964).

Case Two (Pathology Number 12258). This 44-year-old female had developed dull pain in the right cheek two years previously. The pain progressed, and a few months later there was gradual loss of hearing on the right, low pitched tinnitus, and vertigo. About one year prior to admission, the pain and tinnitus increased sharply. Six months later, vision became poor bilaterally. There was no nausea or vomiting.

Physical examination revealed evidence of greatly increased intracranial pressure with choked retinal discs. A ventricular tap confirmed this increased intracranial pressure. Hearing tests revealed a flat sensorineural loss of approximately 45 dB in the right ear (with masking), confirmed by tuning forks. No vestibular tests were performed. At craniotomy, a right cerebellopontine angle tumor, friable, hemorrhagic, and without a capsule, was removed. The VIIth and VIIIth nerves were injured during the operation, and the patient died on the first postoperative day.

The tumor was a large meningioma that had arisen from the surface of the cerebellum. Right temporal bone sections showed that the tumor had grown into the right auditory canal, completely filling this space.
Medulloblastoma

Case (Pathology Number 11620). This 13-year-old female had abruptly stopped growing approximately two and a half years previous to admission. One year later, she began vomiting almost every morning and started having bilateral frontal headaches. One year prior to admission, she began to lose visual acuity, worse on the right. Ten months prior to admission, she began having intermittent vertigo. Later came attacks of hiccoughs lasting two to three hours.

Physical examination revealed bilateral dilated pupils that reacted poorly to light, restricted visual fields and a right hemianopsia, a horizontal nystagmus to the left, and the fundi showed the discs to be pale and cupped. There were dissociated retinal veins. Neurological examination revealed a diminished right corneal reflex, Romberg positive to the right and rear, staggering to the right, and poor finger-to-nose reaction on the right. Hearing tests showed a bilateral flat sensorineural hearing loss of approximately 30 dB on the right and approximately 40 dB on the left. The Weber did not lateralize, and the hearing level was confirmed with forks. Except for the spontaneous nystagmus, no evaluation was made of the vestibular apparatus. An exploratory craniotomy was performed; no resection was attempted. The patient never regained consciousness and died the first postoperative day.

The tumor proved to be a cerebellar glioma which pathologically was termed a medulloblastoma. The temporal bones (Fig 3, A and B) showed the tumor to appear to invade both the VIIth and VIIIth nerves on the right side. Pathological examination, the tumor was found to invade both the VIIth and VIIIth nerves on both sides.

Eosinophilic Granuloma

Case (Pathology Number 11083). This 33-year-old male had a lifelong history of chronic mastoiditis and discharge on the right side. He was admitted to the hospital because of an acute exacerbation and toxicity. On admission, the patient had a temperature of 100.4°F, tender neck nodes on the right, and tenderness over the right forehead and right maxillary antrum. Examination of the right ear canal showed mucopus present in the external canal, and there was a posterosuperior perforation in the tympanic membrane. Visualization of the middle ear through the perforation revealed granulation tissue. Hearing tests on admission revealed a 60 dB conductive hearing loss on the right, but when adequate masking was applied to the left ear, the right ear was found to be non-responsive. This was confirmed with tuning forks. The admitting diagnoses were acute exacerbation of right chronic otitis media with cholesteatoma, acute meningitis, and the possibility of a brain abscess.

The day after admission, a right complete mastoidectomy was performed and cholesteatoma was confirmed. Surgery did not include opening the sigmoid sinus, the inner ear, or going into the internal auditory canal. The patient did well and was discharged the sixth postoperative day. Ten days later, he was readmitted in a comatose state. He had been complaining of pain over the right ear in the interim and expired a few hours after admission.

At autopsy, the middle ear space was seen to contain what was pathologically termed an eosinophilic granuloma, which had extended into the inner ear and had completely filled the internal auditory canal (Fig 4, A and B). There was diffuse pachymeningitis.

Astrocytoma

Case (Pathology Number 12570). This 11-year-old male had begun having nausea and vomiting approximately one month prior to admission. Over the succeeding days, he began having some staggering to the left on walking, drowsiness, generalized headache which would occur every three to four days, and bouts of nuchal rigidity. Approximately two weeks prior to admission, he developed diplopia which lasted for three days. One week prior to admission, a lumbar puncture was performed and there was increased spinal fluid pressure. The family history revealed that a maternal aunt had had a cerebellar pontine angle tumor.

Physical examination revealed vital signs to be normal. Neurological examination showed smell to be decreased. Left pupil was slightly larger than the right and both reacted poorly to light. There was no spontaneous nystagmus, but both fundi revealed disc edema and fundal hemorrhages. The left finger-to-nose test was clumsy, as was the left heel-to-knee. On Romberg testing, he swayed to the left. The audiogram revealed hearing to be normal bilaterally. No vestibular tests were performed. At craniotomy, a pineal tumor was removed and the operation was described as difficult. The patient expired a few hours postoperatively. On pathological examination, the tumor was found to be a glioma and was classified as an astrocytoma. The right temporal bone (Fig 5, A and B) revealed that the tumor had grown into the right internal auditory canal, filling about one-third of the canal. The tumor had not produced any local destruction.

Discussion

A complete microscopic study of the internal auditory canals in 883 temporal bones revealed four small asymptomatic acoustic neurilemmomas, three neurilemmomas which had been operated on, two meningi-
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Fig 3 (A)—Path. No. 11620, Right. Cerebellar medulloblastoma (arrow) that expanded into the IAC and invaded the VIIth and VIIIth nerves. ×11, H & E stain.

Fig 3 (B)—Path. No. 11620, Right. Same temporal bone section as 3(A). The tumor is identical to that seen in the opposite ear. ×125, H & E stain.

Fig 4 (A)—Path. No. 11083. Right middle ear with eosinophilic granuloma invading inner ear. Tympanic membrane and external ear canal are at extreme left. Tumor passes through the oval window (small arrows) and the round window (large curved arrow). The promontory is the bone between the 2 windows. X111, H & E stain.

Fig 4 (B)—Path. No. 11083. Same temporal bone section as 4(A), with histologic pattern at 165× magnification. The tumor appeared to spread to the IAC by way of the neural channels in the spiral ligament.
omas, one medulloblastoma, one eosinophilic granuloma, and one astrocytoma.

As mentioned, the four small acoustic neurilemmomas were considered of sufficient importance to have been reported previously (Leonard and Talbot, 1970), in detail with clinical and pathological findings. Related data from other specimens, including cellular proliferations thought to be possible precursors to neoplasms, were also discussed.

The three postoperative acoustic neurilemmomas are presented here only for complete coverage of the neoplasms found. It is interesting that this study covered a period during which the neurosurgical service at The Johns Hopkins Hospital, headed by Walter Dandy, was especially active. Because acoustic neurilemmomas and other cerebellopontine angle tumors were of special interest to Dr. Dandy, it is a credit to his and his colleagues' skill that more of these patients did not come to autopsy. Obviously, a large number of these patients did die, either immediately postoperatively or at some later time, and a number of the temporal bones ultimately were received in the Otological Research Laboratory. However, in the years covered, the three cases presented were the only ones in which tumor was found in the sectioned temporal bones. The clinical and pathological picture of these three cases is classic.

One autopsy specimen was from a patient who had multiple neurofibromatosis, or von Recklinghausen's disease. He was a 13-year-old male (Pathology Number JHH 15681) who developed an intracranial tumor which proved on exploratory craniotomy to be a neurilemmoma of the third ventricle. The patient died postoperatively, and a complete autopsy was performed. No other central nervous system neumomata were found. Specifically, the entire VIIIth nerves were examined and there was no evidence of acoustic neurilemmoma. The relationship of multiple neurofibromatosis and acoustic neurilemmoma is well established. Hitselberger and Hughes (1964) reported 12 such cases with bilateral acoustic tumors in this syndrome.

Meningiomas were found within the internal auditory canal in two patients; in the first patient the small tumor was discovered accidentally. In the second patient there was a large meningioma in the right cerebellopontine angle. Preoperative diagnosis had been a benign neoplasm, probably a meningioma. It is fairly well accepted that meningiomas originate from the clusters of cells normally present at the tips of the arachnoid villi (Bailey and Bucy, 1931). Arachnoid villi are frequently present in the internal auditory canal, as they are in three other sites within the temporal bone—within the temporal bone—within the region of the geniculate ganglion, and in the sulcus of the greater and lesser superficial petrosal nerves in the roof of the eustachian tube.
The medulloblastoma that was reviewed represents one of the highly malignant gliomas, this one having arisen in the cerebellum and spread to adjacent areas including the internal auditory canal.

The eosinophilic granuloma that was noted is, of course, not a true neoplasm, but is a particular type of chronic inflammatory response. It is a "new growth," and can cause local destruction. This case illustrates this point as there was considerable destruction within the temporal bone, including the inner ear and internal auditory canal. There can be little doubt that this lesion led directly to the death of this patient.

The final case reviewed was that of an astrocytoma. The entire clinical history of the patient included only a month. At operation, the neoplasm was listed as a pineal tumor, and pathologically it was described as an astrocytoma.

The authors are not implying that the cases found in the review of the temporal bone series represents the incidence of these lesions in the general population. These autopsied patients were drawn from an unselected hospital population, but because of the close liaison between the otolaryngological and neurosurgical services, a high percentage of the patients came from the neurological surgery service.

References
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