Surgical removal of giant acoustic neurinomas involving the skull base

Report of two cases

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Neurinomas arising from the peripheral branch of the acoustic nerve distal to the internal auditory canal in the temporal bone are rare. Two advanced skull-base neurinomas are described which were situated mainly in the temporal petrous bone, and extended to the parapharyngeal space anteriorly, to the lateral cervical portion inferiorly, into the sphenoidal sinus medially, and into the middle and posterior cranial fossae compressing the brain stem. Both patients had been deaf for several years without other neurological deficits. The operative findings revealed that the fifth, seventh, and caudal cranial nerves were intact; therefore, it was suspected that these neurinomas originated primarily within the cochlea or the vestibule in the temporal bone. The tumors were completely removed via an extradural approach, with good results. Since the surgical treatment of such advanced skull-base neurinomas is difficult, the operative infratemporal fossa approach is described in detail.

KEY WORDS ● eighth nerve tumor ● acoustic neurinoma ● skull-base tumor ● cranial nerve ● operative technique

ALTHOUGH acoustic nerve tumors arise anywhere along the course of the eighth cranial nerve, most acoustic neurinomas are found within the meatus of the internal auditory canal, producing a cerebellopontine angle syndrome. It is uncommon for an acoustic neurinoma to originate from the distal peripheral branch beyond the internal auditory canal. These rarely act as enlarging tumors of the petrous bone that extend to involve the intracranial cavity and extracranial space.

Access to the skull base remains one of the great challenges for neurosurgeons. Particular difficulty is encountered in treating advanced skull-base tumors that extend into the petrous apex and the infratemporal fossa, which are not easily reached due to interference with the following structures: the temporomandibular joint, the mastication muscles, the internal carotid artery, the internal jugular vein, the facial nerve, the vestibulocochlear organs, and the caudal cranial nerves.

The authors have had experience with two large skull-base neurinomas which extended to the parapharyngeal space, the lateral cervical portion, the middle cranial fossa, and the posterior fossa. The tumors were removed totally using an infratemporal fossa approach with good results.

Summary of Cases

Presentation

Case 1. This 14-year-old boy was referred to our hospital because of deafness for 6 years and developing hoarseness. He had been deaf in the left ear since 9 years of age, but the cause of the hearing disturbance was not known. During the 2 years before admission, he had complained of periodic diffuse headaches, and 12 months later hoarseness developed and grew gradually worse. He was referred to an otologist, who identified a left recurrent nerve palsy and a large skull-base tumor on computerized tomography (CT). The patient was referred to the neurosurgical department of Kin-ikyo Chuo Hospital.

There was no evidence of nevi or subcutaneous nodules associated with neurofibromatosis. A neurological examination disclosed mild sensory disturbance in the
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**Fig. 1.** Case 1. **Upper:** Axial computerized tomography (CT) scans demonstrating a homogeneously enhanced mass arising from the petrous bone and extending into the middle and posterior cranial fossae. The medial half of the petrous bone was destroyed and the remaining lateral portion was elevated upward. The mass occupies the entire carotid canal. Inferiorly, the tumor is located near the parapharyngeal space and extends into the deep cervical portion abutting the axis. The tumor border is well demarcated without evidence of bone invasion. **Lower:** Coronal CT scans showing vertical extension of the tumor. A thin layer of bone surrounding the upper surface of the mass and expanding into the middle fossa suggests that the tumor lies in the extradural space.

left side of the face, sensorineuronal hearing loss of the left ear, mild left vocal cord palsy, diminished pharyngeal reflex on the left side, and moderate weakness of the left sternocleidomastoid muscle. The facial nerve was intact, and taste sensation was normal. There was no paresis of the extremities, the deep-tendon reflexes were normal, and no cerebellar signs were noted. Response to audiometry and caloric testing on the left side was absent. Electronystagmography revealed nothing significant.

Axial CT showed a homogeneously enhancing mass in the left temporal bone with extension toward the middle cranial fossa and the posterior fossa, compressing the cavernous sinus superiorly and the brain stem from the left side. The medial half of the petrous bone was destroyed and the remaining lateral portion was elevated upward. The mass occupied the entire carotid canal and enlarged the jugular foramen; however, the internal meatus on the affected side was intact. Inferiorly, the tumor was located near the parapharyngeal space and in the deep cervical portion abutting the axis. The tumor border was well demarcated without evidence of bone invasion. A coronal CT scan showed vertical extension of the tumor and, since there were thin bone fragments surrounding the upper surface of the mass and expanding into the middle fossa, the appearance suggested that the tumor was situated in the extradural space (Fig. 1). Carotid arteriography demonstrated severe stenosis at the cavernous and the petrous portions of the internal carotid artery with superomedial displacement of the cavernous portion and an inferomedial shift of the petrous section. Vertebral angiography demonstrated that the major blood supply of the left internal carotid artery territory was provided via the left posterior communicating artery. Lateral shift of the petrosal vein and complete obliteration of the jugular bulb were also observed.

**Case 2.** This 18-year-old woman was admitted to Kushiro Rousai Hospital suffering from progressive facial weakness on the left side. She noted left-sided hearing disturbance at the age of 10 years, and she had been deaf in the left ear for 8 years. Six months before admission, she experienced muscle spasm at the angle of the left side of the mouth which ceased spontaneously within several weeks. Four weeks before admission, the left-sided facial weakness recurred and progressed. She was referred to the neurosurgical department for consideration of surgical treatment.

Bone-conduction audiometry demonstrated slightly preserved hearing capacity; however, there was no response to caloric testing. Complete peripheral-type left facial nerve palsy including loss of taste was observed. The fifth and ninth through 12th cranial nerves were not involved. A transoral biopsy of the retropharyngeal mass failed to clarify the histological diagnosis. Radiological findings were very similar to those in Case 1. A CT scan disclosed a well-enhanced mass in the left skull base. The tumor was situated mainly in the petrous bone, destroying the structures of the middle ear and obliterating the eustachian tube. In the middle fossa, the tumor elevated the left cavernous sinus through the foramen lacerum and extended into the sphenoid sinus as far as the midline. The mass entered the posterior fossa through the posterior wall of the petrous...
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FIG. 2. Case 2. Axial computerized tomography scans with contrast enhancement demonstrating a skull-base tumor. The mass extends to the left parapharyngeal space, into the deep temporal fossa, and into the posterior fossa destroying the petrous bone. The lateral half of the petrous bone is elevated and thickened.

Operative Procedure

As a result of the radiological findings in Cases 1 and 2, it was suspected that in both cases the tumor originated from the petrous portion of the temporal bone and was situated extradurally. For an extensive exposure of the temporal area around the auricle, a broad semicircular skin incision was made. The scalp flap was turned, avoiding injury to the frontal and orbital branches of the facial nerve. The external auditory canal was transected at the junction of the bone and cartilage. To prevent exteriorization of the surgical cavity, a blind-sac closure of the external auditory canal was performed. The facial nerve trunk was identified at the retromandibular space and the parotid gland was partially resected to mobilize the parotid plexus of the nerve. The auricular scalp flap was elevated anteroinferiorly over the temporomandibular joint capsule, preserving the subcutaneous great auricular nerve. The zygomatic arch was sectioned and inverted without detaching the masseter or the temporal muscles. The anterior half of the sternocleidomastoid muscle was freed from the underlying mastoid process. To identify the caudal cranial nerves, the internal carotid artery, and the jugular vein, the neck in the retromandibular fossa was dissected. The cicatrized internal jugular vein and the tumor were observed in the deep cervical portion. The temporomandibular joint was disarticulated using a raspator without injuring its capsule to avoid postoperative arthritis, and then the glenoid fossa was drilled away. The mobilized mandibular head was then retracted inferiorly, after which the infratemporal fossa was accessible.

The anterior half of the mastoid process and the tympanic bone were drilled away, anteriorly from the sigmoid sinus and laterally over the facial canal. Following resection of the lateral tympanic bone, the tumor filling the whole tympanic cavity was exposed. The digastric muscle was sectioned and the styloid process was removed with several attached muscles. The facial canal was exposed initially at the stylomastoid foramen by a diamond burr. Although the geniculate ganglion including a part of the horizontal portion of the facial nerve was compressed tightly between the hard tumor and the wall of bone canal, in both cases the nerve was identified and dissected free; however, it was damaged during isolation. Thereafter, skeletonization of the semi-circular canals and the jugular foramen as well as an additional subtotal petrosectomy were performed.

The tumor was removed: first the cervical portion, and then the infra- and intratemporal portions. The tumor was fed by multiple fine arteries branching from the underlying carotid artery; however, bleeding was controllable under direct observation in a wide operative field. The venous bleeding, which occurred when the tumor was dissected from the cavernous sinus, was also easily controlled by packing with Gelfoam. The mass extending into the middle fossa and the posterior fossa through the posterior wall of the petrous bone was carefully dissected from the dura. Total tumor removal was completed extradurally; brain retractors were not required. To prevent a postoperative cerebrospinal fluid leak, multiple tiny tears in the exposed dura were closed with 6-0 nylon sutures under microscopic vision. The seriously damaged segment of the facial nerve was resected. A graft using a cutaneous nerve (the great auricular nerve for Case 1 and the sural nerve for Case 2) was transplanted to bridge the gap between the extradural facial nerve trunk and the intradural nerve root, because the proximal facial nerve was only distinguishable from the acoustic nerve at the cerebellopontine angle cistern.

Surgical Outcome

A postoperative axial CT scan revealed total removal of the tumor in both cases (Fig. 3). The pathological diagnosis was typical schwannoma in each case. The postoperative course of both patients was uneventful.
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the eighth cranial nerve, 2~ and the site of the
tumors as seen in the present two cases are excep-

and they achieved full social recovery. Cerebrospinal
fluid leakage was not observed. Malocclusion due to
drilling away the glenoid fossa did not occur, although
one patient (Case 2) complained of a slight weakness
of mastication for several weeks. The dysfunction of
the caudal nerves in Case 1 gradually improved and his
hoarseness disappeared. Eight months after the opera-
tion functional recovery of the facial nerves began and
continued in both patients. The grafted facial nerve of
Case 1 provided full functional recovery, but that of
the other patient attained only a partial recovery.

Discussion

Acoustic nerve tumors can arise anywhere along the
course of the eighth cranial nerve,20 and the site of the
primary growth in cases where the tumors are small is
confined to the cochlea or to the vestibular labyrinth or
is situated in the internal auditory canal.22 The majority
of acoustic neurinomas arise within the internal audito-
ry canal. Although there are many reports in the
literature of neurinomas arising primarily within the
cochlea and the vestibule in the temporal bone, large
tumors as seen in the present two cases are excep-
tional.1,5,7,19,20,23,34

In considering the origin of neurinomas, several cra-
nial nerves should be excluded: the trigeminal nerve,
facial nerve, and jugular foramen nerve. In addition,
the ascending sympathetic nerve runs along the internal
carotid artery in the petrous bone; however, neurino-
mas arising from such a nerve have not been reported.
In the present cases, the findings on preoperative CT
scans suggested an advanced trigeminal neurinoma;
however, both neurinomas were extradural and were
separated effortlessly from the third division of the
trigeminal nerve at its extradural portion. The caudal
nerves, including the ninth, 10th, and 11th nerves in
the jugular foramen, were also dissected from the tumor
without evidence of any adhesion to those nerves. Fur-
thermore, full functional recovery was observed.

Facial paresis was not evident in the first patient and
both patients had been deaf for several years preceding
the facial nerve symptoms. The participation of the
facial nerve, however, cannot be disregarded. Although
the geniculate ganglion including part of the horizontal
portion of the facial nerve was compressed between the
hard tumor and the wall of the bone canal, the nerve
could be identified and dissected. Additionally, the
greater superficial petrosal nerve was not only identified
but was also unimpaired. When a neurinoma originates
from the tympanic segment of the facial nerve, it usu-
ally presents with a hearing loss due to invasion of the
middle ear, and facial paralysis may occur later.14,21
However, the chorda tympani of the facial nerve was
also observed near the incus in the middle ear. A
neurinoma originating from the stapedial branch can
be excluded based on the anatomical location.

Progressive facial palsy is invariably associated with
facial neurinoma if the neurinoma arises from the
region around the geniculate ganglion.2,9,10,15,17,21 Since
most intratemporal facial neurinomas extend into the
region of the tympanum,13 other symptoms including
ipsilateral conductive or neuronal hearing loss or otitis
media with middle ear extension are often encoun-
tered.15,18 It is less common for these symptoms to
precede a facial palsy,15 and long-term deafness for
several years before the facial symptoms developed has
rarely been reported in cases of facial neurinoma arising
from the geniculate ganglion or horizontal portion; such
tumors usually extend only to the middle or posterior
fossa, with the exception of skull-base invasion.6,12

In a review of 600 temporal bones, Saito and Baxter16
reported five cases of undiagnosed infratemporal facial
neurinomas. Thomsen and Jorgensen22 reported four
cases of undetected acoustic neurinomas, including
three tumors originating in the cochlear branch of the
acoustic nerve, among 150 cases of temporal bone
dissections. Stewart, et al.,19 examined 893 temporal
bones and found five occult schwannomas of the ves-
tibular nerve, including intralabyrinthine cases. Clinical
reports of facial neurinomas in the temporal bone are
much more frequent than those of neurinomas arising
from the acoustic nerve, even in large symptomatic
tumors. With the exception of neurinomas arising from
the internal auditory canal, those arising in the vesti-
bulocochlear branches may be more common than is
generally recognized. On the other hand, in diagnosis of
a large infratemporal facial neurinoma, it appears to
be difficult to exclude the participation of the acoustic
nerve. Based on the evidence discussed it is strongly
suspected that the advanced skull-base tumors pre-
sented in this report are acoustic neurinomas originat-
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ing from the peripheral branch in the temporal bone, which is distal to the internal auditory canal. While the surgical approach to such advanced skull-base neurinomas is still difficult, in both of the patients described total removal was performed by the infratemporal fossa approach with satisfactory results.

The authors would like to emphasize the usefulness of the infratemporal approach for the removal of advanced lateral skull-base tumors, especially those situated in the entire petrous bone.

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References


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