Greater superficial petrosal nerve neurinoma

Case report

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The authors report a case of middle cranial fossa neurinoma arising from the left greater superficial petrosal nerve in a 21-year-old woman who presented with a left-sided otitis media that chronically recurred over a period of 5 years. On examination, the patient had a left-sided mild conductive hearing impairment and a slight disturbance in tear secretion on the left side, with sensory disturbance in the left palate. Three-dimensional computerized tomography scans clearly demonstrated the enlargement of the foramen lacerum and foramen ovale, and heavily T₂-weighted magnetic resonance images revealed the tumor's location along with the course of the greater superficial petrosal nerve and its extension into the tympanic cavity. Following complete surgical excision of the tumor and tympanoplasty via a middle cranial fossa approach, the patient retained useful hearing without facial palsy.

Key Words • greater superficial petrosal nerve • neurinoma • middle cranial fossa • facial nerve

The greater superficial petrosal nerve (GSPN) is a branch of the facial nerve that functions in the innervation of the lacrimal gland and mucous membranes of the nasal cavity and palate. Anatomically, this nerve breaks away at the geniculate ganglion of the facial nerve and courses anteromedially to exit the superior surface of the temporal bone via the facial hiatus. From there it courses anteromedially and continues slightly inferiorly, passing under Meckel's cave toward the foramen lacerum, at which point it joins the deep petrosal nerve from the sympathetic carotid plexus. The two nerves together form the vidian nerve. A neurinoma arising from the GSPN presents as a middle cranial fossa tumor that compresses the temporal lobe and causes a decrease in lacrimation and sensory disturbance in the palate. Neurinomas that definitely arise from the GSPN are extremely rare; only three cases have been reported.3,9,21 Because a large neurinoma arising from the geniculate ganglion of the facial nerve is also located in the middle cranial fossa,1–3,5,8,11,14–20,22 to define whether the origin is the GSPN and/or the geniculate ganglion is difficult. Therefore, it is possible that some tumors that have been reported as originating in the geniculate ganglion may have arisen from the GSPN.

We present a rare case of a GSPN neurinoma located in the middle cranial fossa that was diagnosed on the basis of neurological, radiological, and operative findings, and we discuss reported cases of GSPN or facial nerve neurinoma presenting in the middle cranial fossa.

Case Report

This 21-year-old woman had been treated for chronic recurrence of left-sided exudative otitis media for 5 years. An intracranial mass had been discovered during radiological examinations performed at another hospital, and the patient was admitted to our hospital for surgical treatment in September 1997.

Examination. An admission neurological examination revealed a mild left-sided hearing impairment (right ear 5 dB, left ear 18.8 dB on a pure-tone audiogram) and a mild disturbance in tear secretion on the left side. An electrogustometric examination revealed a mild sensory disturbance in the left half of the soft palate (right side 18.8 dB and left side 34 dB). No loss of taste in the anterior two thirds of the tongue was recorded. Electroneurography of the left facial nerve showed normal function. There was no sensory disturbance in the distribution of the trigeminal nerve.
A computerized tomography (CT) scan revealed an isodense mass in the left middle cranial fossa and bone erosion at the temporal base along the foramen lacerum and at the anterior aspect of the petrous bone with partial calcification of the tumoral rim. The mass was enhanced homogeneously after infusion of contrast medium and appeared to be in contact with the internal carotid artery (ICA) in the temporal bone with extension into the tympanic cavity (Fig. 1 left). Three-dimensional CT scanning demonstrated enlargement of the foramen lacerum and the foramen ovale (Fig. 1 right). A magnetic resonance (MR) image confirmed a tumor at the same location, 2.5 × 2.5 × 2 cm in size, that caused indentation of the temporal lobe (Fig. 2). Heavily T₂-weighted MR images clearly demonstrated the tumor’s location and its extension into the tympanic cavity (Fig. 3). The tumor was located along the course of the GSPN and did not extend along the facial nerve canal. Left carotid artery angiography revealed an avascular mass in the middle cranial fossa.

Operation. After a balloon catheter had been inserted into the left ICA to control bleeding, a left middle cranial
fossa approach was performed. The dura was peeled away from the base of the left middle cranial fossa and the tumor, located extradurally, was recognized. The dura had not been infiltrated by tumor. The pale yellow tumor was soft and contained very few blood vessels. The mass was completely removed piece by piece. The ICA in the petrous bone was recognized after removal of the tumor. The tegmen tympani had been destroyed by the tumor, which extended into the tympanic cavity without adhesion to the ear ossicles. After removal of the tumor, the tegmen tympani was covered with the temporal fascia. In spite of careful tumor removal, it was impossible to identify the GSPN, the otic tube, or the geniculate ganglion of the facial nerve. The origin of the tumor was considered to be the GSPN.

Postoperative Course. Postoperatively, the patient did not exhibit any disturbance in left facial or trigeminal nerve function, although a slight deterioration in hearing (30 dB) due to a build up of exudate was noted on the left side. The histological diagnosis of the tumor proved to be neurinoma, mainly Antoni Type A. On examination 17 months later, only a mild hearing impairment on the left side was demonstrated.

Discussion

Incidence of Facial Nerve Neurinomas

Neurinomas arising from the facial nerve are rare, comprising only 0.8% of all intrapetrous mass lesions. In a series of 527 cerebellopontine angle tumors presented by King and Morrison, there were 13 cases of neurinoma or neurofibromas of the facial nerve in the petrous bone or intracranial cavity. Facial nerve neurinomas involve any segment of the nerve. Among 239 cases reviewed by Lippin, et al., the tympanic segment was most frequently involved (58%), followed by the vertical (48%), the labyrinthine (including the geniculate ganglion) (42%), and the mental segments (30%). Intracranial involvement, such as that including the posterior or middle cranial fossa, was rare (19%).

Because the geniculate ganglion is close to the anterior surface of the petrous bone, it is clear that some neurinomas arising from the geniculate ganglion will be partially located in the petrous bone, with their bulk in the middle cranial fossa. However, they are usually located in the temporal bones, and those located in the middle cranial fossa are rare. To date, there have only been 25 reported cases of facial nerve neurinomas presenting as a middle cranial fossa mass. Among them, three tumors were reported as having only a GSPN origin (Table 1).

Symptoms of Facial Nerve Neurinoma

A precise diagnosis of the level of facial nerve involvement is necessary to locate the tumor in the facial canal. Assessment of facial nerve function by electromyography, stapedial reflex, electrogustometry, and measurement of lacrimal or salivary secretion may be helpful to define the origin and extent of the tumor. Measurement of bilateral tear secretion can disclose any damage to the GSPN. Methods used to determine chorda tympani function include observing impaired or abolished taste by using electrogustometry and measuring salivary secretion from the submandibular glands. In our patient, electrogustometry showed impairment of the left soft palate without impaired taste, which implies dysfunction of the GSPN, although other facial nerve functions were intact. Therefore, our patient showed only GSPN dysfunction among the many functions of the facial nerve both before and after surgery. It was difficult to decide that the origin of tumor was the GSPN, based only on symptoms in the three cases reported previously. Each patient presented with a peripheral facial palsy as an initial symptom, although the diminution of lacrimal secretion was recognized on admission.

A uniform clinical presentation of patients with facial nerve neurinomas originating in the region of the geniculate ganglion consists of facial palsy with hearing loss and typically absence of involvement of other cranial nerves, cerebellar signs, or intracranial hypertension. Among 25 reported cases of middle cranial fossa neurinomas, facial palsy was recognized in 23 cases, although it was preceded by hearing loss in six cases.

The hearing disturbance may be conductive or sensorineural, depending on whether the tumor origin is proximal or distal to the geniculate ganglion. Patients may present with cochlear nerve symptoms in the absence of facial nerve deficit where there is either damage to the inner ear or compression of the cochlear nerve in the meatus, resulting in sensorineural hearing loss or extension into the tympanic cavity with ossicular involvement causing conductive hearing loss. Our patient exhibited conductive hearing impairment as a result of the tumor’s extension into the tympanic cavity; this was diagnosed as chronic otitis media. Thirteen percent of 238 cases were initially diagnosed as “chronic otitis media” because the patients complained of ear pain and otorrhea, which was caused by a mass in the ear canal. Tumor extension into the middle ear was recognized in five of 25 cases of middle cranial fossa neurinoma.
### TABLE 1
Summary of cases with GSPN or facial nerve neurinomas in the middle cranial fossa*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>Symptoms &amp; Signs</th>
<th>Duration</th>
<th>Tumor Origin</th>
<th>Size</th>
<th>Op</th>
<th>Extent of Removal</th>
<th>Postop Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tremble &amp; Penfield, 1936</td>
<td>42, M</td>
<td>FP (com), deafness (partial), tinnitus</td>
<td>5 yrs</td>
<td>lt GSPN</td>
<td>ND</td>
<td>radical mas</td>
<td>total</td>
<td>ND</td>
</tr>
<tr>
<td>Kleinsasser &amp; Friedman, 1959</td>
<td>19, F</td>
<td>FP (com), hearing difficulty</td>
<td>5 yrs</td>
<td>lt GSPN</td>
<td>egg sized</td>
<td>MF exploration</td>
<td>total</td>
<td>no change</td>
</tr>
<tr>
<td>Furlow, 1960</td>
<td>48, M</td>
<td>FP (com), loss of hearing (com), generalized convulsion</td>
<td>13 yrs</td>
<td>rt GSPN</td>
<td>tennis-ball sized</td>
<td>MF exploration</td>
<td>total</td>
<td>no change</td>
</tr>
<tr>
<td></td>
<td>44, M</td>
<td>FP (com), loss of hearing (com), convulsion</td>
<td>12 yrs</td>
<td>rt GG</td>
<td>large</td>
<td>temporal cranio &amp; mas</td>
<td>total</td>
<td>no change</td>
</tr>
<tr>
<td>Curtin &amp; Lang, 1964</td>
<td>20, F</td>
<td>facial paralysis (com), hearing disturbance (partial)</td>
<td>8 yrs</td>
<td>rt tympanic segment</td>
<td>large</td>
<td>radical mas &amp; MF exploration</td>
<td>total</td>
<td>worsened hearing (deafness)</td>
</tr>
<tr>
<td>Hora &amp; Brown, 1964</td>
<td>37, F</td>
<td>hearing difficulty (mild), tinnitus, JP (com)</td>
<td>1 yr</td>
<td>lt horizontal portion</td>
<td>ND</td>
<td>radical mas</td>
<td>subtotal</td>
<td>worsened hearing (deafness)</td>
</tr>
<tr>
<td>Stewart, 1966</td>
<td>38, M</td>
<td>facial paresis (com), decreased hearing (partial), visual blurring</td>
<td>26 yrs</td>
<td>lt side, unknown</td>
<td>lemon sized</td>
<td>frontotemporoparietal crano</td>
<td>subtotal</td>
<td>no change except for visual disturbance</td>
</tr>
<tr>
<td>Pulec, 1972</td>
<td>67, F</td>
<td>hearing loss (com), tinnitus, facial paresis</td>
<td>2 yrs</td>
<td>lt intrapertros</td>
<td>large</td>
<td>MF approach</td>
<td>total</td>
<td>ND</td>
</tr>
<tr>
<td>Isamat, et al., 1975</td>
<td>4, F</td>
<td>hemifacial spasm, deafness (com), tinnitus, FP (com)</td>
<td>7 mos</td>
<td>lt GG</td>
<td>1 × 1.5 cm</td>
<td>MF approach</td>
<td>total</td>
<td>no change</td>
</tr>
<tr>
<td>Liliequist, 1978</td>
<td>42, F</td>
<td>hearing loss (partial), tinnitus, FP (partial)</td>
<td>3 yrs</td>
<td>rt labyrinthine</td>
<td>4 × 4 cm</td>
<td>sub cranio</td>
<td>total</td>
<td>worsened FP (partial) hearing; ND</td>
</tr>
<tr>
<td>Stewart, 1966</td>
<td>19, M</td>
<td>facial paresis (com), loss of hearing (com), headache, tinnitus</td>
<td>12 yrs</td>
<td>lt labyrinthine</td>
<td>huge</td>
<td>ND</td>
<td>subtotal</td>
<td>ND</td>
</tr>
<tr>
<td>Tew, et al., 1983</td>
<td>57, F</td>
<td>hearing difficulty (mod), FP (com)</td>
<td>10 mos</td>
<td>lt GG</td>
<td>2 cm</td>
<td>sub extradural approach</td>
<td>total</td>
<td>ND</td>
</tr>
<tr>
<td>Schneg &amp; de Trivoblet, 1984</td>
<td>28, M</td>
<td>hearing difficulty (mild), FP (com), hearing difficulty (slight), headache</td>
<td>1 yr</td>
<td>lt GG</td>
<td>1.5 cm</td>
<td>sub extradural approach</td>
<td>total</td>
<td>unchanged hearing</td>
</tr>
<tr>
<td>Murata, et al., 1985</td>
<td>39, F</td>
<td>FP (com) &amp; tinnitus, hearing disturbance (mild), headache</td>
<td>13 yrs</td>
<td>rt labyrinthine</td>
<td>4 × 4 cm</td>
<td>sub cranio</td>
<td>total</td>
<td>worsened hearing (deafness)</td>
</tr>
<tr>
<td>Kienzle, et al., 1986</td>
<td>41, F</td>
<td>FP (com), decreased hearing, tinnitus, headache</td>
<td>17 yrs</td>
<td>rt GG</td>
<td>3 cm</td>
<td>sub cranio</td>
<td>total</td>
<td>ND</td>
</tr>
<tr>
<td>Rosenblum, et al., 1987</td>
<td>26, F</td>
<td>loss of hearing, FP (com)</td>
<td>2 yrs</td>
<td>rt GG</td>
<td>2.1 × 1.5 cm</td>
<td>temporal cranio &amp; extradural approach</td>
<td>total</td>
<td>no change</td>
</tr>
<tr>
<td>King &amp; Morrison, 1990</td>
<td>69, F</td>
<td>hearing disturbance (conductive)</td>
<td>2 yrs</td>
<td>rt GG</td>
<td>ND</td>
<td>MF approach</td>
<td>partial</td>
<td>no change</td>
</tr>
<tr>
<td>Symon, et al., 1993</td>
<td>37, F</td>
<td>FP (severe), loss of hearing (com)</td>
<td>5 yrs</td>
<td>GG</td>
<td>1 × 1.4 cm</td>
<td>MF approach</td>
<td>total</td>
<td>improved FP (severe) unchanged hearing</td>
</tr>
<tr>
<td>Dolenc &amp; Korošič, 1996</td>
<td>42, F</td>
<td>facial paresis (mild), mixed loss of hearing (mild), facial weakness (mild)</td>
<td>7 yrs</td>
<td>GG</td>
<td>3 × 4 cm</td>
<td>MF approach &amp; translabyrinthine</td>
<td>total</td>
<td>normal hearing</td>
</tr>
<tr>
<td></td>
<td>44, F</td>
<td>vertigo, FP (com), loss of hearing (com), hearing disturbance (mild), facial sensory disturbance</td>
<td>15 yrs</td>
<td>GG</td>
<td>2 × 2 × 3 cm</td>
<td>MF approach</td>
<td>total</td>
<td>improved hearing</td>
</tr>
<tr>
<td>Yamaki, et al., 1998</td>
<td>46, F</td>
<td>hearing disturbance (mild), FP (mild)</td>
<td>6 mos</td>
<td>lt labyrinthine</td>
<td>2.5 × 2 cm</td>
<td>MF &amp; mastoidectomy approach</td>
<td>total</td>
<td>worsened FP (mod) &amp; hearing (deafness)</td>
</tr>
<tr>
<td>present study</td>
<td>21, F</td>
<td>hearing disturbance (mild)</td>
<td>5 yrs</td>
<td>lt GSPN</td>
<td>2.5 × 2 cm</td>
<td>MF approach</td>
<td>total</td>
<td>useful hearing no FP</td>
</tr>
</tbody>
</table>

* Com = complete; cranio = craniotomy; FP = facial palsy; GG = geniculate ganglion; mas = mastoidectomy; MF = middle fossa; mod = moderate; ND = not described; sub = subtemporal.
† Recurrent facial palsy.
‡ Seventh–12th, seventh–11th cranial nerve, or end-to-end surgical creation of nerve communication by using peripheral nerve graft was performed later in these cases.
§ End-to-end surgical creation of facial nerve communication and tympanoplasty were performed in this case.
Greater superficial petrosal nerve neurinoma

**Radiological Examination**

The correct diagnosis of a facial nerve neurinoma may be made on radiological grounds, provided that the tumor extends some distance along the intrapetrous segment of the facial nerve. The facial nerve canal is enlarged on tomography or on CT scanning.

In our case, three-dimensional CT scanning clearly demonstrated the eroded area of the middle cranial fossa, resulting in an enlargement of the foramen lacerum and foramen ovale. The MR images revealed a homogeneously enhanced mass compressing the temporal base in the middle cranial fossa. The tumor occupied the epidural space from the facial hiatus to the foramen lacerum. It was apparently located along the course of the GSPN and its extension was not related to the facial nerve.

In cases of facial nerve neurinoma in the middle cranial fossa, skull x-ray films and tomograms demonstrated erosion in the anteromedial aspect of the petrous bone and shell-like or curvilinear calcification around the tumor rim. At the geniculate ganglion, where the nerve is very close to the dura of the middle fossa and may even be exposed, the tumor produces erosion into the middle fossa, producing a characteristically smooth defect on top of the petrous bone. This eroded area is the superior surface of the temporal bone near the facial hiatus and was not present in the enlargement of the foramen lacerum shown in our case. Both CT and MR images show a homogeneously enhanced mass compressing the temporal base, and they are essential to diagnose the location of the tumor in the temporal bone. Some authors reported that bone-targeted high-resolution CT scanning is superior to MR imaging, because delineation of the complex bone structures within the petrous bone is important to determine tumor location and extension as well as involved surrounding structures. In our case, the heavily T2-weighted images clearly demonstrated the relationships among the tumor, inner ear structures such as cochlea and labyrinth, cranial nerves in the internal auditory canal, and the brain.

Differential diagnosis of middle fossa masses includes meningioma, neurinoma of the trigeminal nerve, cholesteatoma, glomus tumor, hemangioma, cholesterol granuloma, metastatic tumor, and epidermoid cyst. Meningiomas arising from the floor of the middle fossa would not be expected to erode the petrous bone. Neurinomas of the trigeminal nerve would involve the anteromedial middle fossa and petrous apex rather than its midportion. Epidermoid cysts erode the petrous bone. However, they present a different density or intensity in comparison with neurinoma on CT or MR imaging.

**Surgical Approach**

We selected the middle cranial fossa approach for tumor extirpation. Demonstration of the relationship between the mass and the facial nerve during the operation verified the diagnosis of the tumor’s true origin. The tumor was located along the course of the GSPN, and the ICA in the petrous bone was revealed after tumor removal. Furthermore, the geniculate ganglion as well as the GSPN were not recognized. These findings show that the origin of this tumor was the GSPN.

The surgical approach to remove a facial nerve neurinoma should be selected according to the location and extension of the individual tumor. The middle cranial fossa approach may be selected for tumors of the labyrinthine portion and/or those extending to middle cranial fossa or the petro temporal portion. For tumors of the petro temporal and/or cerebellopontine angle, a suboccipital transmeatal approach may be used. Tumors on the tympanic and vertical portion of the facial nerve can be dealt with via a transmastoid approach. Among the 25 reported cases of facial nerve neurinomas in the middle cranial fossa, the subtemporal or middle cranial fossa approach was selected in 19 cases.

A postoperative complication in cases of facial nerve neurinoma is facial palsy, because surgical extirpation of the tumor results in complete removal of the involved facial nerve. In our case, there was no connection with the geniculate ganglion of the facial nerve, resulting in no facial palsy even after complete removal of tumor. There has only been one other case without facial palsy before and after surgery that has been reported.

Hearing function can be preserved in cases in which there is no tumor involvement in the inner ear or cochlear nerve. Conductive hearing impairment caused by tumor is treated by tumor removal and middle ear reconstruction. In our case, the ear ossicles were not affected by tumor extension into the tympanic cavity; thus useful hearing was preserved following tumor removal and tympanoplasty. Of the 25 described cases of middle cranial fossa neurinoma, there were two in which useful hearing was preserved postoperatively.

**References**


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