Recovery of low-frequency sensorineural hearing loss following resection of a greater superficial petrosal nerve schwannoma

Case report

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Facial nerve schwannomas can occur anywhere from the internal auditory canal to the parotid gland. Schwannomas arising from the greater superficial petrosal nerve are exceedingly rare.

The authors report the case of a 63-year-old woman who presented with a selective low-frequency hearing loss of 3 weeks’ duration. Neurological examination demonstrated a House–Brackmann Grade II facial paresis and asymmetrical hearing loss on the left side. Audiometric evaluation showed a significant loss of low-frequency hearing with a speech reception threshold (SRT) of 30 dB and a speech discrimination score (SDS) of 88% on the left side. Magnetic resonance imaging revealed a 2.4-cm enhancing left middle fossa mass. Near-complete resection was performed via a left temporal craniotomy. The tumor was located in the Glasscock triangle and had invaded the petrous bone overlying the cochlea. A very small piece of the tumor over the cochlea was left in order to preserve hearing. A postoperative audiogram showed significant improvement in the patient’s hearing, with an SRT of 20 dB and an SDS of 100%. The histological findings were consistent with schwannoma.

The patient experienced postoperative improvement of hearing function despite cochlear involvement, which has previously been reported as an unfavorable factor for postoperative hearing outcome in facial nerve schwannomas. (DOI: 10.3171/JNS-07/07/0181)

KEY WORDS • cochlea • facial nerve • geniculate ganglion • hearing loss • petrosal nerve • schwannoma

Schwannomas of the facial nerve can occur anywhere along the nerve from the internal auditory canal to the parotid gland. The labyrinthine, tympanic, and vertical segments are the most common sites.2,6 The main presenting symptom is usually an isolated facial nerve palsy, but hearing loss may also be associated. In such cases, favorable postoperative outcome of hearing function would depend on the presence of a conductive hearing loss and absence of tumor involvement of the inner ear or the cochlear nerve.6

Schwannomas of the GSPN are exceedingly rare, and only a handful of cases have been described in the literature. In this report, we present a case in which the patient presented with a selective low-frequency hearing loss and improvement of this preoperative hearing loss was achieved in spite of the cochlear involvement by the tumor.

Case Report

History and Presentation. This 63-year-old female, a professional pianist, presented with a 3-week history of difficulty in hearing low-frequency sounds on the left side. Upon further questioning she reported experiencing left-eye dryness for the last 30 years and a mild facial asymmetry for the last 6 years. She denied any taste-related problems.

Examination. Her neurological examination revealed a House–Brackmann Grade II facial paresis. Audiometric evaluation showed a significant loss of low-frequency hearing with an SRT of 30 dB and an SDS of 88% on the left (Fig. 1). An MR imaging study demonstrated a 2.4-cm enhancing left middle fossa mass. Near-complete resection was performed via a left temporal craniotomy. The tumor was located in the Glasscock triangle and had invaded the petrous bone overlying the cochlea. A very small piece of the tumor over the cochlea was left in order to preserve hearing. A postoperative audiogram showed significant improvement in the patient’s hearing, with an SRT of 20 dB and an SDS of 100%. The histological findings were consistent with schwannoma.

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Schwannomas of the GSPN are exceedingly rare, and only a handful of cases have been described in the literature. In this report, we present a case in which the patient presented with a selective low-frequency hearing loss and improvement of this preoperative hearing loss was achieved in spite of the cochlear involvement by the tumor.
tumor involvement of the nerve. Because of the patient’s profession, a small amount of the tumor invading the cochlea was left in place in order not to violate the cochlea.

Postoperative Course. The patient’s recovery from surgery was uneventful. An audiogram obtained 6 months after the operation showed significant improvement of the low-frequency hearing loss with an SRT of 20 dB and SDS of 100% on the left side (Fig. 1). The patient’s facial weakness progressed to Grade III in the early postoperative period, but subsequently improved, returning to the baseline (Grade II). Postoperative MR imaging failed to demonstrate any visible enhancing tumor, despite the fact that a

![Image](image-url)
A piece of tumor was left intentionally over the cochlea (Fig. 3). The histological findings were consistent with the diagnosis of schwannoma.

**Discussion**

The GSPN, which arises from the GG, innervates the lacrimal gland and the mucous membranes of the nasal cavity and the palate. It exits the facial hiatus and courses anteromedially along the middle fossa floor forming the medial border of the Glasscock triangle. After joining the deep petrosal nerve of the sympathetic carotid plexus, it exits the skull as the vidian nerve. Schwannomas arising from the GSPN are extremely rare. Since the GG is close to the surface of the petrous bone, schwannomas of the GG may also occur as middle fossa masses, although the majority of these occur inside the temporal bone. Therefore, it may actually be difficult to determine whether a tumor is really of GSPN or GG origin, especially when it is large. In our case, the following two intraoperative findings suggested the tumor to be of GSPN origin: 1) it did not show more proximal involvement than the facial hiatus; and 2) it occupied the Glasscock triangle of the middle fossa floor, where the GSPN would normally run (Fig. 2). Although some authors have been able to distinguish these two neural structures at the time of tumor resection, we and others have not been able to identify them. This factor, however, does not seem to have a significant role in the overall postoperative outcome with respect to the facial nerve.

The hearing loss associated with facial nerve schwannomas, in general, may either be conductive because of the invasion of the tympanic cavity or sensorineural due to the involvement of the inner ear or cochlear nerve depending on whether the tumor is mainly proximal or distal to the GG. In this region, the facial nerve segments are in close proximity to the cochlea. For instance, the average distance between the labyrinthine portion of the facial nerve and cochlea has been reported to be 0.4 mm. The position of the cochlea corresponds to the area medial to the first genu of the facial canal and posterior to the groove of the GSPN, and its apex is located just underneath the proximal groove of the GSPN (Fig. 4). In addition, the average depth of the cochlear apex from the middle fossa floor is less than that of the basal turn of the cochlea due to the downward inclination of the anterior surface of the petrous bone from posterior to anterior. Therefore, a tumor growing from the GSPN would involve the cochlear apex first. Because each segment of the cochlea maps specific hearing frequencies with progressively higher frequencies from the apical to the basal turn, patients with GSPN schwannomas may manifest higher frequency hearing loss early in the course of their disease.
basal turn,7 our patient’s preoperative low-frequency hearing loss corresponded to the anatomical involvement of the cochlear apex by the tumor.

In the present case, the preoperative audiogram showed a low-frequency hearing loss with an air–bone gap. Interestingly, this type of audiogram is more common in conductive hearing loss. It is mostly seen when some form of fixation is present in the sound transmission system showing a stiffness tilt, but it may be also due to an abnormality in the apical part of the cochlea,6 as in our patient. Nevertheless, it can be difficult to distinguish low-frequency conductive hearing loss from low-frequency sensorineural hearing loss because both may produce the same shape in an air conduction audiogram. In the presence of a high bone conduction threshold, the bone conduction measurements may be misleading and may result in a false air–bone gap.6 Overall, the audiogram may be limited in its ability to reveal the status of the apex of the organ of Corti in cases of low-frequency hearing loss with normal high-frequency thresholds.5

Patients with low-frequency hearing loss will manage well if the hearing ability is preserved in the high-frequency range. Therefore, simple speech tests may not reveal this type of hearing loss.5 In our case, the patient’s career as a professional musician not only alerted her at an earlier stage of the hearing loss, but also made it an important goal of the surgery to preserve the hearing. A tiny piece of tumor was intentionally left behind so as to not violate the cochlea and to preserve the hearing. To explain how further improvement of the hearing took place would be somewhat speculative. One possible explanation could be that removing the bulk of the tumor mass relieved the pressure over the cochlea, which was being transmitted through the osseous defect.

Conclusions
Schwannoma of the GSPN is an exceedingly rare tumor and to our knowledge an association with a selective low-frequency hearing loss has not been documented in the literature previously. Another unique feature of this case is the postoperative improvement of the patient’s hearing despite the involvement of the cochlea by the tumor, which has previously been reported as an unfavorable factor for the outcome of hearing function in facial nerve schwannomas. We believe that leaving a small piece of tumor over the cochlea led to the functional preservation, which has become an increasingly important concept in current neurosurgical practice.

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References

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