Hearing Improvement after Resection of a Large Jugular Foramen Schwannoma: Case Report

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ABSTRACT

Although hearing improvement after surgery for small tumors of the cerebellopontine angle has been reported, the mechanism by which surgery leads to the improvement in hearing remains controversial. We report a patient who sought treatment for progressive tinnitus and hearing loss. Magnetic resonance imaging showed a large (5-cm) schwannoma in the cerebellopontine angle. At surgery the lesion was found to originate from rootlets of cranial nerve X at the jugular foramen. The patient underwent gross total resection of the tumor. Immediately after surgery, his hearing improved dramatically. We believe that our patient represents an example of hearing impairment at least in part referable to direct compression of the brainstem. Importantly, the patient's hearing deficit was completely reversible. Some authors claim that surgery to preserve hearing may be contraindicated in patients with speech discrimination scores below 50%. However, when extrinsic brainstem compression may contribute to the cause of such a hearing decrement, postoperative improvement in hearing may be a reasonable expectation.

KEYWORDS: Jugular foramen schwannoma, hearing, retrosigmoid craniotomy, cerebellopontine angle

The rate of hearing preservation after surgery for vestibular schwannomas in patients with serviceable hearing varies. For small (< 2 cm) tumors, the reported rates of preserved hearing range from 25 to 47%. Hearing improvement after surgery for tumors involving cranial

nerve (CN) VIII is an uncommon but well-documented event. Nevertheless, most surgeons consider severe hearing loss (typically defined as either pure-tone average greater than 50 dB or a speech discrimination score of less than 50%) to be at a level that is not useful for communication,

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and, in fact, may hinder understanding of speech in the better ear.

Tumors of the jugular foramen often manifest with tinnitus or hearing loss. Such a presentation in isolation, as well as growth of the schwannoma purely intracranially, is unusual. The cause of hearing loss associated with tumors other than those arising from CN VIII may differ from that of hearing loss associated with intrinsic tumors of CN VIII.

We present a 28-year-old man with a schwannoma of CN X associated with isolated tinnitus and severe hearing loss that completely reversed after surgery. The location of this lesion within the cerebellopontine angle (CPA) suggests that the patient's hearing loss, which was related to mass effect, may be distinguished from that caused by intrinsic CN VIII involvement. Specifically, we propose that this case may illustrate selective speech discrimination loss related to the compressive effect of the tumor on integrative neurons within the brainstem or spiral ganglion afferents. We further propose that the hearing loss was reversible because the brainstem compression was ameliorated by surgery.

CASE REPORT

A 28-year-old man, who had a history of Ewing's sarcoma of the lumbar spine 16 years earlier, sought treatment for long-standing tinnitus and progressive loss of hearing in the right ear. On his initial consultation, he complained that he was unable to communicate by telephone with the affected ear. Earlier, the patient had also undergone amputation below the knee and a colostomy related to complications from treatment of the Ewing's sarcoma. Magnetic resonance imaging (MRI) showed a large (4.9- × 4-cm) schwannoma within the lateral cerebromedullary fissure centered at the CPA. The tumor extended to the tentorium superiorly and to the jugular foramen inferiorly (Fig. 1).

On physical examination, the patient had grossly diminished hearing on the right. His face was symmetric and without numbness, his palate was elevated symmetrically, and his tongue was midline without evidence of atrophy. There were no other significant cranial nerve findings. His motor strength was 5/5. A finger-to-nose

Figure 1 (A) Axial and (B) coronal gadolinium-enhanced magnetic resonance images show a 4.9- × 4-cm tumor within the cerebellopontine angle.

Figure 2 (A) Axial and (B) coronal postoperative magnetic resonance images confirm gross total resection of the tumor.

examination showed mild ataxia of the right upper limb. Despite the patient's prosthesis, his gait was normal, without impaired tandem walk. The patient was evaluated with a preoperative audiogram. Pure-tone hearing averages were mildly impaired at 13 dB. Speech reception threshold was 30 dB, and the patient's speech discrimination score for the right ear was 8% at 30 dB above threshold.

The patient underwent an image-guided retrosigmoid craniotomy for resection of the tumor. Intraoperative stimulation of the facial nerve and monitoring of the auditory brainstem response were used as surgical adjuncts. At surgery the lesion was found to arise from the jugular foramen. Tumor entered the internal auditory canal (IAC), with CN VII/VIII draped over the dome of the tumor. The IAC was drilled out, allowing for tumor removal. Gross total resection was obtained (Fig. 2).

Postoperatively, the patient noted immediate improvement in the hearing of his right ear. On postoperative day 3, his speech discrimination score was 100%. The patient developed vocal cord paralysis requiring Gelfoam[®] (Pharmacia and Upjohn, Kalamazoo, MI) medialization and percutaneous gastrostomy, but his hospital course was otherwise unremarkable.

DISCUSSION

Sensorineural hearing loss can be the result of injury at any level of the auditory pathway, from the organ of Corti to the central ascending auditory pathways. Clinically, it makes sense to categorize most hearing loss as either cochlear or retrocochlear. Cochlear deafness may best be treated with surgical strategies that address the underlying etiology of cochlear deafness, such as cochlear implantation. Retrocochlear causes of deafness are typically associated with tumors of the auditory nerve or CPA.

However, lesions throughout the brainstem and cortex have been reported to cause deafness. For example, deficits in sound localization have been associated with a cavernous malformation compressing the nucleus of the trapezoid body.¹ Bilateral injury of the inferior colliculi has been associated with word deafness while perception of pure-tone averages was preserved.^{2,3} In general, as the location of a lesion ascends along the auditory pathway, the manifestation of the deficit becomes subtler, reflecting early cross-connections between the right and left sides of the auditory pathway. These subtle deficits reflect the integrative role of auditory nuclei as auditory information is processed from the cochlear nuclei to the cortex.

Our patient had a relatively selective loss of speech discrimination, which was completely reversed after his schwannoma was resected with concomitant decompression of his brainstem. We believe that the relative sparing of pure-tone audition in our patient is consistent with compression of brainstem nuclei. Within the cochlear nucleus, for example, broadly tuned neurons such as octopus cells of the posterior ventral cochlear nucleus play an integrative role by responding as physiological "on"-type units that are likely to be prominently involved in the physiological processing of speech.⁴ In addition, the efferent fibers of the olivocochlear pathway may play a role, by selective inhibition of cochlear resonance, in integrating nontonal speech information that became deranged by the compression.

Although in large series of patients undergoing attempted hearing preservation surgery for acoustic neuroma neither pure-tone averages nor speech discrimination scores were found to be a better prognostic predictor of hearing preservation postoperatively,⁵ hearing improvement after resection of acoustic neuromas has been previously reported,^{6,7} and in these reports the hearing parameter most likely to improve has been the speech discrimination score. However, these reports failed to discriminate between patients who had purely intracanalicular tumors and those with significant compression of the brainstem.

Multiple etiologies exist to account for hearing loss attributable to tumors of the CPA. Traditionally, hearing loss associated with the presence of a mass originating within the IAC has been attributed to the deleterious effects of direct compression of CN VIII, invasion of the axons of the primary cochlear afferents, and impairment of the blood supply to the nerve and cochlea.^{5,8} Thus, hearing loss associated with even small tumors arising within the IAC is thought likely to represent damage to CN VIII itself. In these cases, improvement in speech discrimination following tumor resection likely reflects the role in speech discrimination subserved by primary afferent (and/or olivocochlear) fibers of CN VIII as opposed to integrative neurons within the brainstem per se.

In the present case, the tumor originated at the level of the jugular foramen and extended rostrally toward the tentorium; only a small amount of tumor extended into the porus acusticus. Although the nerve was draped over the tumor, the extent of IAC involvement was minimal compared with the large compressive effect of the tumor on the brainstem. Nevertheless, the role of decompression of CN VIII cannot be completely disregarded.

Finally, we did not test the patient's preoperative auditory brainstem evoked response or otoacoustic emissions. Other authors have proposed that the pattern of distortion of these evoked waveforms may suggest the location of injury within the auditory pathway.^{9,10} Qui et al argue that preserved otoacoustic emissions in the presence of ≥ 35 -dB sensorineural hearing loss likely reflects a blockade of ascending sensory input at the level of the superior olivary complex (i.e., a retrocochlear hearing loss). Similarly, we propose that brainstem compression may significantly contribute to decrements in speech discrimination scores, and that the effect of compression is more likely to affect this measure than the pure-tone average. Unfortunately, we did not assess our patient for otoacoustic emissions; in patients with severely impaired speech discrimination, brainstem compression, and relative sparing of pure-tone thresholds, such preoperative testing would be valuable to determine whether the severely impaired speech discrimination scores not only correlated with impaired brainstem responses, but also with a better prognosis for hearing preservation (or even improvement) after surgery.

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Commentary

The authors report a large nonacoustic cerebellopontine angle (CPA) neuroma, which

was removed with remarkable recovery of speech discrimination. They speculate that this is due to removal of pressure on the brainstem with reactivation of more central auditory pathways. Although interesting, there are few data to support this theory. The cochlear nerve was also stretched over the top of the tumor and could have been a second site of compression along with the brainstem. Auditory brainstem audiometry was not performed so there are no data to support the site of lesion; therefore, I believe this theory must remain speculation.

This case does show the importance of attempting hearing preservation in nonacoustic CPA tumors. Primary epidermoids of the CPA are notorious for producing poor speech discrimination out of proportion to pure-tone loss. Often, speech discrimination will recover when these tumors are removed. Removal of meningiomas may also result in hearing improvement. The overall success rate for removal of nonacoustic CPA tumors is superior to results with similar-sized acoustic neuromas. The authors are to be congratulated for an excellent outcome with this very large tumor.

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