Temporal Bone Paraganglioma: Hearing Outcomes and Rehabilitation

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Abstract

Despite numerous advancements in the treatment of skull base tumors, the management of jugular paragangliomas remains controversial and nuanced. Contemporary treatment objectives focus not only on achieving durable tumor control but also on maximizing post-treatment quality of life. While this philosophy places appropriate attention on limiting cranial neuropathies, less discussed morbidities such as hearing loss can have a profound impact on quality of life outcomes. The objective of this retrospective review is to provide a comprehensive review of hearing outcomes following the treatment of tympanojugular paragangliomas. A particular focus is placed on the variable audiologic outcomes as it relates to disease pathology, extent of disease, as well as chosen treatment modality. This review underscores the importance of considering treatment-related effects on hearing.

Keywords

- ▶ paraganglioma
- ▶ glomus tympanicum
- ▶ hearing outcomes
- ▶ stereotactic radiosurgery

Introduction

Since the first reported surgical resection in 1945, the treatment of jugular paragangliomas has remained controversial.¹ In the decades that followed these initial treatment efforts, several classification systems were proposed in hopes of standardizing surgical practices. Unfortunately, the inherent heterogeneity of the collective pathology prevented such schematics from being universally adopted. Variations in size, growth rate, and anatomic location have precluded a one-size-fits-all approach to treatment. Instead, contemporary strategies have focused on designing individualized plans utilizing microsurgical resection, stereotactic radiosurgery (SRS), combination therapy, and, with increasing frequency, active surveillance. While the merit of each therapeutic modality continues to be debated, treatment objectives have remained remarkably consistent. Today, the goal of modern-day management is to achieve durable radiographic and clinical tumor control while simultaneously preserving critical neurovascular structures.

Limiting treatment-related sequela can be challenging given the intimate anatomic relationship between neoplasm and the adjacent structures of the surrounding skull base.

The breadth of potential complications has been widely reported, with particular attention paid to the functionally devastating effects of lower cranial nerve (CN) injuries, namely CN IX to XI. Dysphagia, dysphonia, tracheostomy dependence, and failure to liberate from enteral feedings have all been reported.^{2,3} With the debilitating effects of these aforementioned complications taken into consideration, hearing outcomes can be understandably deprioritized when considering the potential morbidity of various therapeutic options. This emphasis on lower CN neuropathies, along with the relative rarity of jugulotympanic paragangliomas, contributes to the scarcity of literature on pathology-specific post-treatment hearing outcomes.

With the relative paucity of available data, audiologic outcomes from more commonly encountered skull base lesions have become useful surrogates to frame the potential effects of paraganglioma treatment. For instance, in acoustic neuroma patients, postoperative hearing loss (HL) has been identified as an independent predictor in quality of life outcomes.⁴ In fact, a large retrospective review of postsurgical acoustic neuroma patients found that HL—and not facial paresis, disequilibrium or tinnitus—was perceived as the most disabling of all incurred morbidities. While these findings are not directly translatable

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to paraganglioma practices, they underscore the potential impact of hearing conservation efforts in the treatment of skull base lesions. Even when this knowledge is taken into consideration, hearing preservation usually remains of subordinate importance to tumor exposure, internal carotid artery (ICA) control, and lower CN preservation. However, universal disregard of post-treatment hearing outcomes is a grave disservice to the patient. The objective of this article is to provide a review of treatment-related audiologic outcomes for paragangliomas while also discussing potential rehabilitation options for select patients.

Temporal Bone Paragangliomas

From an anatomic perspective, temporal bone paragangliomas can be broadly classified into two groups: tympanomastoid and jugulare paraganglioma tumors. Tympanomastoid neoplasms originate from the middle ear and spare the jugular vein. Meanwhile, jugulare paragangliomas arise from the jugular vein and are capable of spreading to the structures of the adjacent skull base (► **Figs. 1 and 2**). These groupings can be further stratified into specific staging systems. Currently, there are two main grading systems used to stage temporal bone paragangliomas: the Fisch and Mattox and the Glasscock and Jackson. The main difference between the two systems is that the Glasscock and Jackson system grades jugulare and tympanomastoid paragangliomas separately. Conversely, the Fisch and Mattox system uses one scale to grade both disease processes along a single continuum. Familiarity with both systems is helpful when reviewing the literature, as particular authorships often preferentially stratify lesions based on one specific grading scheme.

Tympanomastoid Paragangliomas

Tympanomastoid paragangliomas, also commonly known as glomus tympanicum tumors, arise from rests of paragangliar tissue contained in the middle ear space. Although variations have been reported, these tumors most commonly arise from the medial wall of the middle ear cleft along the course of Jacobson's nerve. Given their anatomic predisposition, it is perhaps unsurprising that patients most commonly present with audiologic disturbances, namely HL and pulsatile tinnitus. Hearing loss can be either sensorineural (SNHL), mixed (MHL), or conductive (CHL). SNHL is rare, often unrecoverable, and related to cochlear invasion and fistulization. CHL, which is significantly more common, is often caused by ossicular mass loading, ossicular discontinuity, as well as tympanic membrane or round window obstruction. Such situations present the unique opportunity to provide durable tumor control, preservation of vital structures, and *improvement* to pre-existing HL. Overwhelmingly, these tumors are treated via surgical removal. However, SRS has been performed and can be offered in select patients with significant comorbidities and unfavorable preoperative risk stratification.

Surgery

As previously alluded to, the majority of paraganglioma literature focuses on durability of tumor control. As such, hearing outcomes are reported infrequently with inconsis-

tent standardization of the reported data. Since CHL is the most common cause for hearing impairment, the reported outcomes are more of a testament to the capabilities and potential of ossicular chain reconstruction than a reflection of the functional impact of the disease process. One of the largest reviews by Patnaik et al stratified hearing-related outcomes based on tumor stage (Fisch and Mattox) as well as surgical technique.⁵ The results were somewhat expected, as postoperative hearing outcomes worsened as the class increased. While patients with class A lesions demonstrated improvements in air conduction levels, those with Class B lesions did not improve. This is unsurprising when one considers that the study standardized treatment such that Class B lesions which were treated by subtotal petrosectomy with middle ear obliteration. A second retrospective review by Forrest FA & Jackson GC et al stratified patients by Glasscock and Jackson classification, while assessing hearing outcomes based on ossicular chain involvement.⁶ Of the 57 patients with postoperative audiograms, only one patient had a 15 dB or greater drop in pure tone average. Patients with an intact ossicular chain improved postoperatively, but had an air bone gap (ABG) with a mean of 16 dB. Surprisingly, the ABG was smaller for patients undergoing ossicular chain reconstruction, with a mean ABG of 13 dB.

Radiation

As previously stated, the treatment of jugulotympanic paragangliomas is primarily surgical. However, select series have been published describing long-term treatment results from SRS.⁷ The goal of treatment in these cases is to arrest tumor growth without the expectation for tumor shrinkage. With this in mind, hearing improvement cannot be realistically expected to improve following treatment. Studies have confirmed this suspicion by reporting post-treatment hearing outcomes in binary fashion. In one such study, of the 36 patients who presented with HL, there were no patients with improvements in hearing. Hearing deterioration was not reported.⁷

Jugular Paragangliomas

Jugular paragangliomas are the most common tumor of the jugular foramen. They arise from rests of paraganglia tissue in the adventitia overlying the jugular bulb. While the close proximity to the surrounding skull base puts lower CNs at risks, large-scale reviews found that the most frequent presenting symptoms were audiologic disturbances. HL and tinnitus were experienced by 77.3 and 71.7%, respectively, while the most common manifestation of lower CN neuropathy was hoarseness, found in 26.4% of patients.⁸

Surgery

Postoperative hearing outcomes are largely a reflection of tumor size and surgical approach. Early surgical efforts popularized by House and Histleberger employed a translabrynthine/transcochlear dissection.⁹ This approach was acoustically ablative and left individuals with no serviceable hearing in the operated ear. Advances in surgical approach came not as a response to hearing conservation, but rather to

Fig. 1 Magnetic resonance imaging (MRI) of left jugular paraganglioma: In this coronal T1-weighted MRI, there is a mass filling lesion of the left jugular foramen.

Fig. 2 Microscopic otoscopy: This is the corresponding physical exam from the same patient as ►Fig. 1. On examination of the left ear, there is a hypotympanic vascular mass behind an otherwise well-aerated middle ear.

Fig. 3 Surgical resection of the lesion demonstrated in ►Fig. 1. A canal wall up mastoidectomy was performed with retrofacial dissection to the jugular bulb.

improved surgical exposure. These calls were answered by the introduction of the infratemporal fossa (ITF) approach by Fisch in the late 1970s.¹⁰ Designed to treat large and intracranial tumors, the radical transtemporal approach provided wide surgical exposure for gross total tumor resection. However, the technique required the middle ear apparatus lateral to the stapes to be removed and the external ear canal over closed via blind sac closure. Such efforts resulted in an expectant maximal CHL. While the ITF approach is still commonly used today, contemporary surgical decision making in the treatment of jugular paragangliomas has become more nuanced. Considerations of subtotal versus gross total resection have transformed the notion of respectability from a binary assessment to a spectrum of ablative efforts.

In the decades that followed the introduction of Fisch's ITF approaches, the focus of paraganglioma treatment shifted from achieving total tumor resection to an increased emphasis on the functional outcomes of postsurgical patients. In the midst of this renaissance, Jackson et al presented what he would describe as "hearing conservation" surgery for jugular paragangliomas tumors.¹¹ The authors assumed that with the exception of translabyrinthine/transcochlear dissection or intracochlear invasion, hearing should be preserved in most patients. Thus, the challenge was to preserve the lateral acoustic conduction system sacrificed within the ITF approaches, without forfeiting surgical exposure. To achieve this, the authors employed an extended infratympanic facial recess that allowed for both preservation of the external auditory canal (EAC) while simultaneously providing adequate exposure for distal ICA control (►Fig. 3). The group established several criteria to assess surgical candidacy. First, the tumor must be confined to the infralabyrinthine chamber (Glascoc and Jackson I–III; Fisch and Mattox B and C). Second, the tumor must be dissectible from middle ear and cochlea. Third, the tumor must not extend anterior into the ITF. Finally, exposure must be sufficient to allow distal control of the ICA. Above all else, the authors emphasize that hearing conservation was an ancillary consideration compared with the importance of distal ICA control and tumor exposure. Their sentinel study described hearing conservation surgery for 38 jugular paragangliomas tumors. Successful conservation of hearing was directly related to tumor size with 83.5% of tumors being Glascoc and Jackson I or II, and 81% classified as Fisch and Mattox B or D. In these instances, a modification of the ITF approach may leave the external auditory canal intact which will reduce the risk of the resultant CHL.

Radiation

HL from lateral skull base radiation can be related to post-radiation changes to the external auditory canal, tympanic membrane, ossicles, cochlea, cochlear nerve, brain stem, and central auditory pathway. Thus, treatment-related HL can manifest as CHL, SNHL, or MHL.¹² Despite the numerous locations for potential audiometric compromise, one of the principal concerns in temporal bone radiation is cochlear toxicity and damage. Conventional external beam radiation was first introduced as a primary treatment method for jugulare paraganglioma tumors in the 1950s. While tumor

control rates were reasonable, the required dose provided significant toxicity to the surrounding neurovascular structures including the cochlea. SRS was subsequently introduced in response to these findings, offering durable tumor control with dramatic improvements in associated morbidity. Patel et al produced the largest study to date examining the post-treatment hearing outcomes from 85 patients with pre-treatment serviceable hearing (American Academy of Otolaryngology Head and Neck Surgery Class A or B) treated with SRS.¹³ The authors demonstrated a low risk of cochlear toxicity over the length of the study. Seven patients developed nonserviceable hearing, which tended to manifest within the first 2 years following treatment. The median bone conduction pure tone average threshold increase was 1.2 dB HL per year. This accompanied a 0% change per year in speech discrimination.

Other Otologic Outcomes

Pulsatile tinnitus is classically cited as a common presenting symptom for temporal bone paragangliomas. Unfortunately, much like hearing outcomes, little is reported on how this symptom responds to therapeutic intervention. Patel et al found that in patients presenting with pulsatile tinnitus, 60% experienced mild-to-moderate improvement in symptom control after SRS treatment.¹³ Similar rates of improvement have been demonstrated in other studies. Dobberpuhl et al retrospectively examined 10 patients with jugular paragangliomas treated by SRS, 8 of which experienced improvement or resolution of pulsatile tinnitus.¹⁴ The study did not identify any individuals whose symptoms worsened. Unfortunately, there is a limited amount of data regarding post-surgical tinnitus improvement.

Hearing Rehabilitation

Hearing Amplification

After undergoing surgical resection with preservation of the external auditory canal, patients may suffer from sensorineural or mixed HL and benefit from hearing amplification devices. These patients should be referred to the appropriate audiology practice for evaluation and discussion of these rehabilitation options.

Bone Anchored Hearing Aid

In patients with single sided deafness or maximal CHL as a result of their treatment, a bone anchored hearing aid (BAHA) remains a viable option. Patients with a subtotal petrosectomy and blind sac closure of the ear canal will have a CHL that will be amenable to a BAHA. These patients with maximal CHL tend to report greater satisfaction with usage of BAHA as it restores a significant amount of binaural hearing. Several options for the BAHA are now available including a percutaneous and a transcutaneous version. A complete discussion on the topic can be found in the section dedicated to bone conduction devices featured in this issue.

Cochlear Implantation

Cochlear implantation remains the only reliable method of auditory rehabilitation for patients with severe to profound

SNHL. In patients with temporal bone paragangliomas, this situation can arise through several causes. Though SNHL is encountered in select postsurgical patients, these situations are most often the result of ablative cochlear dissection, intracochlear erosion secondary to tumor expansion, invasion of the internal auditory canal, or effacement of the eighth nerve in the cerebellopontine angle. In this regard, postsurgical patients are infrequently cochlear implantation candidates. However, in select postsurgical patients with progressive postoperative SNHL and concomitant cochlear preservation, cochlear implantation remains a viable option. Cochlear implantation has been reported in a young patient treated surgically for bilateral paragangliomas. The implanted side was managed via canal wall down mastoidectomy without blind-sac closure. While he initially performed well, his post-operative course was remarkable for electrode extrusion through the tympanic membrane and radiographic evidence of bony resorption of all cochlear turns on the implanted side.¹⁵ While subtotal petrosectomy with blind sac closure has been reported as a successful methodology in patients with chronic suppurative otitis media, this technique has not been reported in patients with paraganglioma.

While cochlear implantation is a relatively infrequent option for postsurgical patients, it has become a viable for patients with radiation-induced SNHL. Successful cochlear implantation has been reported in patients having undergone SRS for acoustic neuroma.^{16,17} Recently, successful cochlear implantation has also been reported in post-radiated paraganglioma patients as well.¹⁸ With the feasibility of implantation in the radiated temporal bones no longer in question, the main focus has shifted to identifying appropriate candidates.

Regardless of the etiology of SNHL, continued radiologic evaluation is a consideration in all patients who are undergoing evaluation of cochlear implantation following treatment of temporal bone paragangliomas. Current recommendations suggest magnetic resonance imaging of up to 3.0T can be performed with indwelling cochlear implants. Nonetheless, distortion artifact is present. Fortunately, this artifact does not extend to the jugular foramen, and thus does not preclude continued radiographic surveillance in patients who have undergone cochlear implantation.¹⁹

Conclusions

Temporal bone paragangliomas are benign slow-growing, hypervascular tumors. Treatment of these indolent lesions has evolved dramatically over the past 80 plus years. Despite the innumerable advancements in surgical technique, SRS, and auditory rehabilitation, the basic tenants of paraganglioma treatment have remained unchanged: hearing should not supersede the primary objective of durable tumor control with preservation of neurovascular function. However, in select patients, there now exists several options and techniques capable of maximizing post-treatment auditory outcomes and with it improving quality of life.

Conflict of Interest

None.

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