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Hearing Improvement after Radiation Therapy for a Facial Nerve Schwannoma: Report of a Case and Review of Literature

Isabella L. Pecorari[1,2] Sayak R. Ghosh[1,2] Howard S. Moskowitz[3,4] Vijay Agarwal[1,2]

[1] Department of Neurological Surgery, Albert Einstein College of Medicine, Bronx, New York, United States
[2] Department of Neurological Surgery, Montefiore Medical Center, Bronx, New York, United States
[3] Department of Otorhinolaryngology, Albert Einstein College of Medicine, Bronx, New York, United States
[4] Department of Otorhinolaryngology, Montefiore Medical Center, Bronx, New York, United States

Address for correspondence Vijay Agarwal, MD, Montefiore Medical Center, The University Hospital for the Albert Einstein College of Medicine, 3316 Rochambeau Avenue, Bronx, NY 10467, United States (e-mail: vagarwal@montefiore.org).

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Abstract

Introduction While facial nerve schwannomas are considered benign, they can impart various significant clinical effects due to pressure on nearby cerebrovascular structures within the cerebellopontine angle (CPA). Although surgical resection and/or radiation therapy often provide definitive treatment of such tumors, posttreatment hearing loss is a common finding. In this report, we present the case of a patient with a facial nerve schwannoma successfully treated with radiotherapy with resultant hearing improvement, an extremely rare clinical finding.

Case Presentation A 63-year-old woman presented with a 1-year history of progressively worsening hearing loss and tinnitus. Brain imaging demonstrated an enhancing lesion of the right CPA measuring $2.7 \times 2.1 \times 3.1$ cm. Pretreatment audiometry evaluation revealed sensorineural hearing loss in the right ear with a pure-tone average (PTA) of 74 dB, speech threshold (ST) of 75 dB, and speech discrimination (SD) of 0%. The patient proceeded with attempted surgical resection, aborted due to significant facial nerve stimulation, and ultimately underwent radiation therapy (50.4 Gy, 28 fractions). At the 1-year follow-up visit, the patient reports subjective hearing loss resolution with PTA of 34 dB, 30 dB ST, and 88% SD on audiological evaluation.

Conclusion Although radiation therapy for schwannomas within the CPA has historically been associated with hearing loss, fractionated stereotactic radiotherapy (FSRT) may provide improved clinical outcomes compared with high-dose radiosurgery. Given the effectiveness of this treatment modality and improved quality of life offered to patients over surgery, FSRT may be considered an initial management option for patients with facial nerve schwannomas.

Keywords
- facial nerve schwannoma
- hearing improvement
- radiation therapy
- CPA mass

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Introduction

Cerebellopontine angle (CPA) tumors constitute a significant proportion of intracranial neoplasms, representing up to 10% of tumors within the brain.[1] Located anterior to the cerebellum, lateral to the pons, and posteromedial to the petrous temporal bone, the CPA is traversed by several cranial nerves and important neurovascular structures.[2] As a result, lesions in this region can impart various clinical effects, including hearing loss, tinnitus, vertigo, and facial pain, among others.[3] Out of the tumors that arise within the CPA region, schwannomas account for the majority of lesions.[1,4]

Schwannomas are benign tumors arising from neural crest-derived Schwann cells, which encase peripheral nerves.[5] As a result of their relatively slow growth, patients may present with gradual symptom onset prior to diagnosis.[6] Although vestibular nerve schwannomas are the most prevalent subtype, schwannomas of the facial nerve are exceedingly rare and can occur anywhere along the extracranial, intracranial, or intra-temporal portions of the seventh cranial nerve.[7]

Current management options for CPA schwannomas include surgical intervention, radiosurgery, radiotherapy, or observation.[8] When deciding between these options, it is important to recognize the potential clinical side effects that patients may experience. Hearing loss is one particular adverse outcome, with over 50% of patients experiencing a decline in hearing after 5 years of radiation therapy.[9] Significant hearing impairment after surgery has also been documented, with some studies reporting hearing loss rates greater than 80% after surgical intervention; however, variability in outcomes can occur based on the surgical approach taken, tumor size, and surgeon experience.[10] Despite the different treatment options available, the decision to pursue specific therapies is dependent on several patient and tumor characteristics.[11,12]

In this report, we present the case of a facial nerve schwannoma that experienced significant hearing improvement after radiation therapy. This is a rare clinical finding given the high rates of hearing decline that is most often documented after radiation to tumors within this region of the brain.[13] Therefore, this case highlights the potential for radiation therapy to offer improved clinical outcomes for tumors within the CPA.

Case Presentation

A 63-year-old woman with a past medical history significant for hypertension presented to otolaryngology clinic with 1 year of progressively worsening right-sided hearing loss and constant tinnitus. During this time, the patient experienced intermittent headaches but denied dizziness, otalgia, or otorrhea. The patient had no family history of hearing loss or history of prior head trauma. On physical exam, vestibulo-ocular reflexes in response to rapid horizontal head impulses revealed a refixation saccade on head impulse to the right. Aside from the right-sided hearing loss, the remaining physical exam was unremarkable.

Initial audiometry evaluation revealed bilateral sensorineural hearing loss, worse on the right. The right ear had a pure-tone average (PTA) of 52 dB (measured on 500, 1000, 2000, and 4000 Hz), speech threshold (ST) of 40 dB, and speech discrimination (SD) of 68% (►**Table 1**). The PTA, ST, and SD of the left ear were 24 dB, 20 dB, and 100%, respectively. Magnetic resonance imaging (MRI) of the internal auditory canal demonstrated an enhancing lesion of the right CPA measuring $2.7 \times 2.1 \times 3.1$ cm. At this time, all treatment options were fully discussed, including surgery, radiation, and observation, including their potential risks and benefits.

The patient was subsequently lost to follow-up until presenting to clinic 14 months later with new-onset vertigo and progressive worsening of right-sided hearing loss. Repeat physical exam was unchanged compared with initial presentation. Audiometry report demonstrated progressive hearing loss of the right ear, with a PTA of 74 dB, 75 dB ST, and 0% SD. MRI revealed an interval increase in size of the right CPA mass, measuring $3.2 \times 2.3 \times 3.4$ cm with extension into the interval auditory canal (►**Fig. 1**). The mass was noted to exert significant mass effect on the pons, right middle and inferior cerebellar peduncles, and right cerebral peduncle, and to cause hydrocephalus due to compression of the fourth ventricle.

A decision was made after extensive discussion of all the options available, including surgical resection, radiation, and observation, to pursue surgical intervention due to the increasing size of the mass and worsening clinical symptoms. A right suboccipital stereotactic craniotomy was initiated. After initial exposure, the tumor was found to stimulate at 0.2 mA over the majority of the tumor, indicating a likely facial nerve schwannoma. A small area of the tumor was able to be safely sent for pathological diagnosis, which confirmed a schwannoma. The surgical resection was subsequently aborted to prevent facial nerve damage after the confirmation of a likely facial nerve schwannoma. The patient recovered from surgery well with no neurological deficits and was

Table 1 Comparison of pretreatment and posttreatment audiometry reports (right ear)

	Speech audiometry		Tone audiometry (Hz)				
	ST (dB)	SD (%)	500	1000	2000	4000	PTA (dB)
Initial diagnostic report	40	68	45	43	48	73	52
Preoperative report	75	0	83	80	60	73	74
First post-RT report	30	80	35	38	43	58	43
Second post-RT report	30	88	20	33	30	55	34

Abbreviations: PTA, pure-tone average; RT, radiation therapy; SD, speech discrimination; ST, speech threshold.

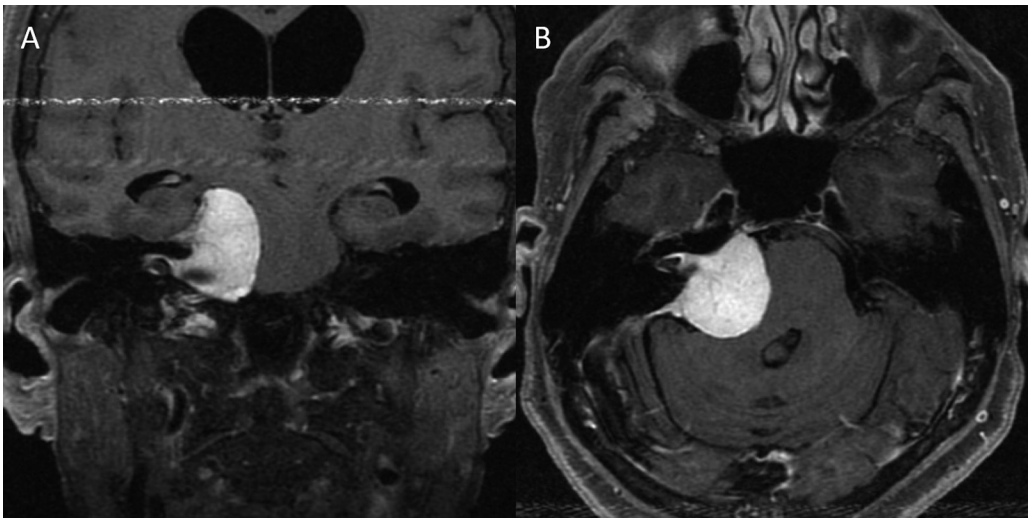

Fig. 1 Preoperative coronal (**A**) and axial (**B**) postcontrast T1-weighted fluid-attenuated inversion recovery magnetic resonance imaging.

noted to have full facial function bilaterally (Grade I House–Brackmann bilaterally). The patient was discharged on postoperative day 4.

At the patient's 2-month postoperative follow-up visit, minor subjective hearing improvement was reported. No wound-healing issues were noted. Due to the size of the lesion, the decision was made to treat it with fractionated radiation. Radiation therapy was initiated with a total of 50.4 Gy delivered in 28 fractions. At the 3-month postradiation follow-up visit, significant improvement in hearing and tinnitus was reported. The patient continued to endorse intermittent dizziness, imbalance during ambulation, and slight transient right-sided facial pain anterior to the ear. Repeat audiometry evaluation at the 6-month postradiation follow-up visit revealed hearing improvement in the right ear, with a PTA of 43 dB, 30 dB ST, and 80% SD. Four months later, the patient demonstrated further improvement in hearing loss with a PTA of 34 dB, 30 dB ST, and 88% SD. Repeat MRI showed the lesion measured $2.9 \times 3 \times 3$ cm, unchanged in size. At the most recent follow-up visit, 1-year after radiation therapy completion, the patient continues to experience amelioration of her right-sided hearing impairment.

Discussion

In this report, we present the case of a patient with a right-sided facial nerve schwannoma experiencing symptoms of hearing loss, tinnitus, vertigo, and facial pain. After undergoing biopsy and subsequent radiation therapy, the patient experienced significant hearing improvement compared with pretreatment audiology reports, with no significant change in size of the lesion. At this time, there is a lack of published cases illustrating improved hearing after radiation, with the majority of studies demonstrating worsened clinical outcomes after such treatment. The results of this case, however, provide evidence that improved hearing outcomes are possible for patients with facial nerve schwannomas treated with fractionated stereotactic radiotherapy (FSRT).

When determining the appropriate treatment of choice for patients with tumors in the CPA, it is critical to make an accurate diagnosis of the type of lesion present. Due to the similar location of facial nerve and vestibular schwannomas, these two entities can be challenging to distinguish preoperatively. Although both tumors have comparable imaging characteristics and may at times be indistinguishable, facial nerve schwannomas have been reliably found to exhibit extension into the genicular ganglion and/or labyrinthine portion of the facial nerve canal.[14] Aside from distinguishing between types of CPA schwannomas, other lesions within this region of the brain must be effectively ruled out from the differential diagnosis prior to treatment initiation. For example, neurofibromas may resemble schwannomas as both are peripheral nerve sheath tumors with overlapping imaging characteristics. However, schwannomas tend to be located peripheral to the nerve, have more clearly defined margins, and demonstrate potential cystic components.[15] Another mass that can arise in the genicular ganglion is facial nerve hemangioma. Like neurofibromas, they can be distinguished from facial nerve schwannomas by the presence of irregular borders as well as a characteristic "honeycomb" appearance on high-resolution computed tomography.[14] While utilizing imaging characteristics is a helpful tool in differentiating tumor types within the CPA, intraoperative findings and pathology are often required to make a definitive diagnosis, especially when needing to distinguish facial nerve from vestibular schwannomas.

Treatment options for patients with facial nerve schwannomas include surgical resection, radiation therapy, or observation. The decision to pursue each modality is dependent on several factors, including symptom severity, tumor size, patient age, the presence of comorbid conditions, and patient preference.[11,12] While radiation therapy and surgery are the main definitive treatments for facial nerve schwannomas, observation alone may be an option for certain patients. Individuals who have medical comorbidities, advanced age, or small and/or asymptomatic tumors, for example, may opt for delayed treatment. Furthermore, patients with only mild

hearing loss and/or normal facial nerve function with a House–Brackmann score of I or II may forego treatment.[14] In these situations, patients are monitored with frequent surveillance imaging to track progressive tumor growth. However, many patients who undergo an initial period of observation may eventually require more definitive treatment interventions. In studies comparing surgical excision to conservative management, anywhere from 19 to 90% of patients managed conservatively eventually underwent a surgical procedure, ranging from exploration to wide decompression and tumor debulking.[16,17] This variability in outcomes may reflect the need for a more robust treatment algorithm for facial nerve schwannomas, bolstered by case data such as the one presented here.

The primary goal of surgical intervention for facial nerve schwannomas is tumor removal with preservation of facial nerve function and hearing.[18] Surgical options include gross total or partial resection, with or without nerve grafting.[19] Exact surgical approach depends on the location of the tumor and includes infratemporal fossa, translabyrinthine, and retrosigmoid approaches, among others.[20] Often, the facial nerve cannot be preserved, in which case grafting may be performed to provide patients with the potential for facial nerve function improvement.[14] In a study of 50 facial nerve schwannoma cases, 76% of patients who received surgical treatment experienced a decline in facial nerve function following surgery, compared with 17% in patients who were offered stereotactic radiosurgery (SRS) and 0% in patients who were observed.[17] The difference in adverse outcomes, however, may reflect a selection bias as patients who underwent surgical management were likely more often to have larger, faster-growing, and more invasive tumors or to have failed prior conservative treatment.

In contrast to surgery, radiation therapy has been considered a relatively safe alternative and is often preferred as an initial treatment in patients with facial nerve schwannomas who have small symptomatic tumors ($<1\ cm^3$), an inability to undergo surgery, or as an adjuvant treatment postsubtotal resection.[17,21,22] In addition to providing a less invasive treatment approach, radiation therapy has been found to be a comparable and possibly superior treatment option with regard to clinical outcomes and quality of life. In a meta-analysis by Rotter et al consisting of 519 patients with facial nerve schwannoma, rates of facial nerve function improvement were similar when comparing surgery to SRS (23 vs. 20%).[19] In comparison to surgically treated patients, those treated with SRS had higher rates of preserved facial nerve function and lower rates of worsening facial nerve function after treatment. However, a selection bias may exist for the surgical treatment of more aggressive and larger tumors, potentially limiting the validity of studies comparing such treatment modalities.

At this time, radiation therapy can be delivered in two forms: FSRT, given as multiple doses over several sessions, or SRS, which provides a single dose of radiation.[23] One study conducted by Shi et al reviewed eight patients with facial nerve schwannomas, seven of which were treated with FSRT. On presentation, six of the patients had facial nerve dysfunction, with House–Brackmann scores ranging from I to IV, and five patients had hearing impairment. After radiation treatment, five patients were noted to have stable tumor size, whereas three patients experienced volume reduction. Furthermore, six of the patients had improvement in clinical symptoms.[21] In another study by Nishioka et al, among four patients with facial nerve schwannoma treated with FSRT, none had worsening of their neurological symptoms, and two patients had reduction in tumor size.[24] These studies, in addition to the present case, demonstrate the safety and potential efficacy of radiation treatment in improving clinical outcomes of patients with facial nerve schwannomas.

In addition to FSRT, single high-dose radiation surgery has also been utilized as a treatment modality for patients with facial nerve schwannomas. In a multicenter study of 42 patients receiving gamma knife surgery, a type of SRS, 37 patients remained neurologically stable or were observed to have clinical improvement on most recent follow-up after treatment, whereas 5 experienced permanent neurological decline, which included worsened hearing loss, facial spasms, and facial weakness.[22] Furthermore, a lower dose of radiation, less than or equal to 12.5 Gy, and an initial tumor volume of less than $1\ cm^3$ were both associated with favorable neurological outcomes. Tumor control was achieved in 90% of patients at 28 months, and survival was 90% at 5 years. In another study of six patients who received gamma knife radiosurgery for facial nerve schwannoma, three patients experienced tumor regression after treatment, whereas the rest were found to have stable tumor volumes.[25] Notably, all patients were found to have preserved facial function after treatment. Another study of 14 patients with facial schwannoma reported shrinkage of tumor volume after radiosurgery in 10 patients, 9 of which experienced either improvement or preservation in their facial nerve function after treatment.[26] Notably, in both of these studies, there was no tumor progression, and hearing loss was relatively unchanged. Fezeu et al presented a total of six series of facial nerve schwannomas treated with SRS, totaling 44 patients, reporting tumor control in 93% of cases, hearing preservation in 95%, and facial nerve deterioration in 5%.[27] Although larger sample sizes could demonstrate stronger statistical significance, this preliminary data demonstrates the efficacy and safety of radiation therapy for treating patients with facial nerve schwannomas.

At this time, data comparing outcomes of FSRT versus SRS for facial nerve schwannoma is currently unavailable. However, comparison of these treatment modalities has been performed for vestibular schwannomas. In a study of 125 patients with vestibular nerve schwannomas receiving either FSRT or SRS, rates of tumor control and preservation of cranial nerve function were similar between treatment groups. However, hearing preservation was found to be 2.5-fold higher in patients receiving FSRT.[28] A systematic review of 19 case studies of vestibular schwannoma that were treated with either FSRT or SRS demonstrated that rates of subsequent treatment interventions were comparable between radiation groups.[29] Additionally, patients treated with either SRS or FSRT had comparable rates of hearing

deterioration after treatment (49% for SRS and 45% for FSRT). Of note, only 2 of the 19 case studies analyzed outcomes from FSRT, indicating a need for more data from this treatment modality. As demonstrated by these studies, significant variability in hearing outcomes exists between these two treatment options. In general, younger patient age, lower radiation doses, more favorable pretreatment hearing levels, and smaller tumor size have all been found to be associated with greater rates of hearing preservation after radiation therapy.[30–32] Interestingly, the patient in this report did not have several of these prognostic indicators, suggesting that other patient characteristics or tumor factors may be responsible for patients experiencing favorable clinical outcomes after radiation.

With regard to survival outcomes, SRS and FSRT have been found to have comparable progression-free survival rates among patients with vestibular schwannomas, with several reports documenting a 5-year tumor control rate of greater than 90% for both radiation modalities.[29,33,34] This is similar to tumor control rates among patients who underwent gross total surgical resection of their CPA schwannomas. Since limited data exist comparing different treatment modalities for facial nerve schwannomas, it may be appropriate to extrapolate expected treatment outcomes from studies on vestibular schwannomas to facial nerve schwannomas until more studies focused on this specific tumor entity are reported.

Conclusion

Radiation therapy to tumors within the CPA often results in long-term posttreatment hearing deficits. In the current case, however, FSRT was shown to result in hearing improvement from a nonserviceable to serviceable level, which is an uncommon clinical finding. This report may indicate that compared with single high-dose radiation therapy, FSRT may be a preferred first-line treatment modality for many patients with CPA schwannomas. Despite the potential risks that may be associated with radiation, a trend toward nonsurgical management is becoming preferred by patients, most likely due to the decreased invasiveness of this procedure and more favorable side effect profile compared with surgery. As a result, further work investigating specific patient characteristics, tumor qualities, and radiation dosing schedules that result in improved clinical outcomes is warranted.

Conflict of Interest

None declared.

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