Gamma Knife radiosurgery for facial nerve schwannomas: a multicenter study

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OBJECT Facial nerve schwannomas (FNSs) are rare intracranial tumors, and the optimal management of these tumors remains unclear. Resection can be undertaken, but the tumor's intimate association with the facial nerve makes resection with neurological preservation quite challenging. Stereotactic radiosurgery (SRS) has been used to treat FNSs, and this study evaluates the outcome of this approach.

METHODS At 8 medical centers participating in the North American Gamma Knife Consortium (NAGKC), 42 patients undergoing SRS for an FNS were identified, and clinical and radiographic data were obtained for these cases. Males outnumbered females at a ratio of 1.2:1, and the patients' median age was 48 years (range 11–76 years). Prior resection was performed in 36% of cases. The mean tumor volume was 1.8 cm³, and a mean margin dose of 12.5 Gy (range 11–15 Gy) was delivered to the tumor.

RESULTS At a median follow-up of 28 months, tumor control was achieved in 36 (90%) of the 40 patients with reliable radiographic follow-up. Actuarial tumor control was 97%, 97%, 97%, and 90% at 1, 2, 3, and 5 years postradiosurgery. Preoperative facial nerve function was preserved in 38 of 42 patients, with 60% of evaluable patients having House-Brackmann scores of 1 or 2 at last follow-up. Treated patients with a House-Brackmann score of 1 to 3 were more likely to demonstrate this level of facial nerve function at last evaluation (OR 6.09, 95% CI 1.7–22.0, $p = 0.006$). Avoidance of temporary or permanent neurological symptoms was more likely to be achieved in patients who received a tumor margin dose of 12.5 Gy or less (log-rank test, $p = 0.024$) delivered to a tumor of ≤ 1 cm³ in volume (log-rank test, $p = 0.01$).

CONCLUSIONS Stereotactic radiosurgery resulted in tumor control and neurological preservation in most FNS patients. When the tumor is smaller and the patient exhibits favorable normal facial nerve function, SRS portends a better result. The authors believe that early, upfront SRS may be the treatment of choice for small FNSs, but it is an effective salvage treatment for residual/recurrent tumor that remain or progress after resection.

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FACIAL nerve schwannomas (FNSs) are rare tumors accounting for 1.9% of all intracranial neuromas and 0.8% of intrapetrous tumors.¹ FNS can arise from any segment of the facial nerve, starting at its origin in the cerebellopontine angle and extending to its ex-

tracranial components.² While such neuromas can mimic the radiographic features of the more common vestibular schwannoma, patients with FNS typically present with signs and symptoms of facial nerve dysfunction.

The optimal management strategy for FNS patients

ABBREVIATIONS FNS = facial nerve schwannoma; GKRS = Gamma Knife radiosurgery; NAGKC = North American Gamma Knife Consortium; SRS = stereotactic radiosurgery.

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remains to be defined. Management options include resection, stereotactic radiosurgery (SRS), fractionated radiation therapy, and observation. Despite improvements in microsurgical techniques, preservation of existing facial nerve function remains challenging.

After SRS for vestibular schwannomas, the 10-year rate of tumor control is typically greater than 90% and the incidence of facial nerve palsy is $\leq 1\%$.^{4–6} Hearing preservation 5 years after SRS is maintained in 60%–70% of these patients, although long-term hearing preservation may be lower.^{5,10–12} As such, given the success of SRS for vestibular schwannomas, its application for FNS seems a natural extension.¹⁵ During the past decade, there have been a few retrospective studies related to FNS management by SRS, and most of these case series have been comprised of small numbers of patients.^{13–16} The goal of this study was to report a multiinstitutional experience with Gamma Knife (Elekta AB) radiosurgery (GKRS) for intracranial FNS.

Methods

Patient Population

Eight medical centers participating in the North American Gamma Knife Consortium (NAGKC) obtained individual institutional review board approvals for participation in this study. A total of 42 patients were identified with intracranial FNS managed at least in part by GKRS. At each center, retrospective clinical outcome analysis of GKRS-treated cases of FNS was performed. The following centers contributed data for this study: the University of Pittsburgh (14 patients); Cleveland Clinic (7 patients); the University of Pennsylvania (1 patient); National Yang-Ming University in Taipei, Taiwan (1 patient); Université de Sherbrooke (6 patients); Yale University (5 patients); New York University (3 patients), and the University of Virginia (5 patients).

The records of FNS patients who underwent GKRS between 1988 and 2014 were evaluated by clinicians at each center for study inclusion. A database with appropriately selected variables was created and sent to the participating centers. Participating centers reviewed the medical records of their patients, entered the data in the spreadsheet, and deleted all patient identifiers from the database. Pooled and de-identified data were screened by an independent third party for errors. Any uncertainties or ambiguities in the data were addressed to the respective contributing center. Afterward, data were transmitted to the NAGKC's clinical trials coordinator to verify compliance with protection of patient information.

Patients were included in the study if they had a histologically diagnosed FNS. Also, patients were included in this study if they had clinical and neuroimaging features consistent with a benign FNS. Features included a medical history absent of prior cancer and an intracranial tumor located along one or more segments of the facial nerve with MRI and/or CT imaging features most in keeping with an FNS. The neuroimaging features included avid contrast enhancement, rare tumor calcification, and bony widening of the facial nerve canal (Fig. 1). The presence of enhancement along the course of the facial nerve (e.g., around the

geniculate ganglion) after intravenous administration of a contrast agent was used to help diagnose an FNS. Similarly, bony erosion around the geniculate ganglion and/or otic capsule or enlargement of the facial canal were other features considered consistent with an FNS. For inclusion, patients were required to have some evaluable neuroimaging and/or clinical follow-up after GKRS.

Patient and Tumor Attributes

The median age of the 42 patients was 48 years (range 11–76 years). The group included 23 (55%) male patients and 19 (45%) female patients. None of the patients had prior radiotherapy. Fifteen patients (36%) had prior surgical resection with histologically confirmed facial nerve schwannomas. The remaining patients displayed neuroimaging and clinical features consistent with a benign FNS. Such clinical features included a predominance of the signs and symptoms related to facial nerve dysfunction (as opposed to vestibular dysfunction).

The mean duration of follow-up was 42 months (median 28 months, range 1–139 months). Preradiosurgical patient characteristics, presentations, and tumor characteristics are detailed in Table 1.

Radiosurgical Technique

The various Gamma Knife Models U, B, C, 4C, or Perfixion (Elekta AB) were used depending on the technology available at the time of radiosurgery for the participating centers. The radiosurgery began with the application of the Leksell Model G stereotactic frame (Elekta AB) using local anesthetic augmented with additional sedation as needed. After stereotactic frame placement, a high-resolution, stereotactic MRI examination was performed. In cases in which MRI was not feasible or when MRI distortion was a concern, a stereotactic CT scan was performed. Thin-sliced axial and/or coronal plane images were obtained after intravenous administration of a contrast agent. Radiosurgery dose planning was then performed by a neurosurgeon in conjunction with a radiation oncologist and medical physicist. All patients were treated with single-session radiosurgery. A typical Gamma Knife dose plan for a patient included in this series is shown in Fig. 1.

The mean volume of the FNSs was 1.8 cm³ (median 0.79 cm³, range 0.04–8.7 cm³). The mean prescription dose delivered to the tumor margin was 12.5 Gy (range 11–15 Gy). The mean prescription isodose line was 52% (median 50%, range 50%–87%) (Table 2). Most of the dose plans involved a multi-isocentric technique; the mean number of isocenters used was 11 isocenters (range 1–30). Dose selection was based on an empirical algorithm that included considerations related to tumor volume, preexisting neurological deficits, proximity to critical structures such as the brainstem, and history, if any, of prior fractionated radiation therapy.

Clinical and Neuroimaging Follow-Up

Clinical and neuroimaging evaluations were generally performed at follow-up intervals of 6 months for the first 2 years after radiosurgery. Facial nerve function was evaluated by using the House-Brackmann grading system,¹⁶ and hearing function was assessed with the Gardner-Robertson

FIG. 1. An example of MR images from a Gamma Knife dose plan with the prescription isodose shown.

classification.¹⁷ For patients without detailed House-Brackmann or Gardner-Robertson assessments, facial nerve and hearing functions were classified as stable, improved, or worsened by the treating clinicians.

In patients who demonstrated no evidence of tumor growth and absence of new neurological findings, follow-up intervals were gradually increased to once every 1–2 years. Whenever feasible, patients underwent evaluations, including neurological examination and neuroimaging, at the respective treating center. However, since all participating institutions represent tertiary referral centers, in some cases follow-up evaluations were performed by the patients' local physicians. In all instances, clinical notes and neuroimaging studies were reviewed by the treating clinicians who performed the GKRS. The follow-up images were compared with the images obtained at the time of GKRS. Tumor dimensions were assessed in the axial, sagittal, and coronal planes. An increase of more than 10% of the planned treatment volume (PTV) or tumor occurrence outside the PTV was deemed tumor progression. Tumor regression was defined as a decrease of the tumor by more than 10%. The tumor was deemed stable if its dimensions were within $\pm 10\%$ of the size at the time of GKRS. Tumor control was defined as regression or stability of the tumor.

Statistical Analysis

Data are presented as median or mean and range for continuous variables and as frequency and percentage for categorical variables. The Wilcoxon signed-ranks test was used to compare the House-Brackmann score just prior to and at the last follow-up evaluation after SRS.

Transient and/or permanent neurological symptoms after SRS were evaluated using a Kaplan-Meier methodology. Patients who were neurologically asymptomatic following SRS were not included in the analysis of postoperative symptom changes. A Kaplan-Meier plot was used to compare the time to development of the neurological symptoms as a function of the tumor volume and radiosurgical margin dose to the tumor.

TABLE 1. Characteristics of patients with facial schwannomas treated with GKRS*

Characteristic	Value
No. of patients	42
Sex	
Male	23 (55%)
Female	19 (45%)
Prior resection	15 (36%)
Age at GKRS (years)	
Median	48
Range	11–76
Side	
Right	21 (50%)
Left	21 (50%)
Follow-up (months)	
Median	28
Range	1–139
Tumor type	
Solid	31 (74%)
Cystic	2 (5%)
Mixed	9 (21%)

* Values represent numbers of patients (%) unless otherwise indicated.

Progression-free survival was analyzed using the Kaplan-Meier method. Also, we analyzed tumor control, House-Brackmann grade stability/improvement, and Gardner-Robertson class stability/improvement at the last follow-up, using the logistic regression analysis to predict the clinically important prognostic factors, including age at the time of SRS, margin dose to the tumor, volume of the tumor with a cutoff point of 1 cm³, preoperative House-Brackmann grade, and preoperative Gardner-Robertson class.

Statistical computation was conducted using IBM SPSS 21. All statistical analyses were 2-tailed, and a p value < 0.05 was deemed statistically significant.

Results

Radiological Outcome

The mean follow-up was 42 months (median 28 months, range 1–138.7 months). Reliable radiographic follow-up was available for 40 of the 42 FNS patients. At last follow-up, the FNS volume was stable in 21 patients (52% of evaluable patients), decreased in 15 patients (38%), and progressive in 4 patients (10%). Thus, the overall rate of local tumor control was 90% at last follow-up.

TABLE 2. Gamma Knife radiosurgical parameters

Attribute	Mean	Range
Tumor volume (cm ³)	1.8	0.04–8.7
Margin dose (Gy)	12.5	11.0–15
Maximum dose (Gy)	24	15–30.4
Prescription isodose (%)	52	50–87

Kaplan-Meier analysis demonstrated the radiographic progression-free survival at 1, 2, 3, and 5 years to be 97%, 97%, 97%, and 90% respectively (Fig. 2). In those patients who had prior surgical intervention, the radiographic progression-free survival rate was 100% at 1, 3, and 5 years (i.e., at each of these time points). For those treated with upfront SRS, the progression-free survival rates at 1, 3, and 5 years were 95%, 95%, and 83%, respectively. There was no statistically significant difference in radiographic progression-free survival between those treated after a prior resection and those treated with upfront SRS (log-rank test, $p = 0.25$).

Clinical Outcomes

Some degree of clinical follow-up information and, in particular, overall neurological function was available in 42 patients. Detailed House-Brackmann grades were available for 40 patients at the time of GKRS and 37 at last follow-up; Gardner-Robertson classes were available for 35 patients at the time of GKRS and 28 at last follow-up (Table 3). At last clinical follow-up, 37 patients were noted to be neurologically stable or improved, but 5 patients had experienced some degree of permanent neurological decline. Throughout the follow-up period of the study, 10 patients (25%) developed various types of either transient ($n = 5$) or permanent ($n = 5$) neurological symptoms following SRS. Those symptoms included intermittent facial spasms, worsened hearing, increased facial weakness, progressive balance impairment, or dizziness.

A Kaplan-Meier plot was used to compare the time to symptomatic manifestations following SRS with respect to the margin dose delivered to the tumors. Figure 3 indicates that a margin dose less than or equal to 12.5 Gy was significantly correlated with an outcome free of new

FIG. 2. Kaplan-Meier plot of tumor progression-free survival after SRS for all FNS patients. The patient numbers along the x-axis represent the number of patients at risk for each major time milestone after radiosurgery.

TABLE 3. Distribution of patients by House-Brackmann grade and Gardner-Robertson class prior to GKRS and at last follow-up

Scale & Grade/Class	No. of Patients (%)	
	Pre-GKRS (%)	Post-GKRS (%)
House-Brackmann grade		
I	9 (23)	11 (30)
II	7 (18)	11 (30)
III	13 (33)	4 (11)
IV	5 (13)	7 (19)
V	3 (8)	1 (3)
VI	3 (8)	3 (8)
Gardner-Robertson class		
I	13 (37)	7 (25)
II	9 (26)	7 (25)
III	7 (20)	5 (18)
IV	3 (8.5)	2 (7)
V	3 (8.5)	7 (25)

* Pre-GKRS and post-GKRS House-Brackmann grades were available for 40 and 37 patients, respectively. Pre- and post-GKRS Gardner Robertson classes were available for 35 and 28 patients, respectively. The sum of individual percentages for each category is not exactly 100% due to rounding.

or progressive neurological symptoms (log-rank test, $p = 0.024$). Twenty-five patients (60%) harbored a tumor with a volume of 1 cm³ or less. In univariate analysis, the patients with a tumor of 1 cm³ or less in volume fared better neurologically (i.e., no new or worsened neurological deficits) than those whose tumor volume was larger than 1 cm³ (Fig. 4, log-rank test, $p = 0.01$). Of note, the status of previous resection (i.e., whether a patient had prior resective surgery or not) had no relationship with an outcome free of new or progressive neurological symptoms after SRS (log-rank test, $p = 0.96$). The small sample size precluded a meaningful multivariate analysis.

Any change in a patient's House-Brackmann grade after SRS was significantly related to a permanent change in House-Brackmann grade at last follow-up on the Wilcoxon signed-ranks test ($p = 0.026$). Furthermore, logistic regression analysis revealed that the maintenance of a House-Brackmann grade ranging from I to III at last follow-up was intensely correlated with a House-Brackmann grade of I to III prior to SRS (OR 6.09, 95% CI 1.7–22.0, $p = 0.006$).

Favorable Outcome After SRS

We defined a favorable outcome after radiosurgery as tumor stability or regression in combination with improved facial and auditory function at last follow-up. At last follow-up, tumor progression occurred in 4 patients (9.5% of all 42 patients or 10% of the 40 patients for whom reliable radiographic follow-up was available), worsening of auditory function in 9 patients (21% of all patients), and worsening facial nerve function in 4 patients (9.5% of all patients) (Table 4). In the univariate analysis, a tumor volume of 1 cm³ or less was associated with a favorable outcome following radiosurgery (OR 5.4, 95% CI 1.29 to 22.6 in logistic regression analysis, $p = 0.021$).

FIG. 3. Kaplan-Meier plot illustrating the significant correlation between a margin dose ≤ 12.5 Gy and an outcome free of new or progressive neurological symptoms after SRS (log-rank test, $p = 0.024$).

FIG. 4. Kaplan-Meier plot showing that patients with a tumor of 1 cm³ or less in volume fared better from a neurological standpoint after radiosurgery as compared with those whose tumor volume was larger than 1 cm³ (log-rank test, $p = 0.01$).

Post-SRS Management

Following SRS, 5 patients received a course of corticosteroid treatment. The corticosteroids were used to treat worsening hearing in 3 patients and facial nerve symptoms in 2 patients. In all 3 cases, the medical treatment improved the symptoms.

An additional patient developed a tumor-related cyst and underwent aspiration of the cyst following SRS. Finally, 1 patient underwent repeat SRS for tumor progression. The repeat SRS was performed 60 months after the initial SRS. The remaining 2 patients with tumor progression after SRS have been observed to date.

Other Serious Adverse Effects

In the current study, there was no evidence of radiation-induced tumor formation or malignant transformation of an existing FNS.

Discussion

The Diagnosis and Natural History of FNS

Facial nerve schwannomas are rare and typically slow-growing tumors. They often present with an insidious onset of minor facial nerve symptoms, and patients may initially be misdiagnosed as having Bell's palsy. FNSs are frequently diagnosed in middle-aged patients. Facial nerve dysfunction is the cardinal feature of these tumors. While normal facial nerve function has been reported in 27% to 54% of FNS patients, a meticulous physical examination and history taking can often reveal some level of facial nerve dysfunction.^{10,21,23} In a recent meta-analysis of 427 cases of FNS, facial weakness (occurring in 63% of cases) and hearing loss (51%) ipsilateral to the tumor were

the most common symptoms.²⁴ Hemifacial spasm and facial weakness were the most frequent symptoms when the FNS arose in the cerebellopontine angle.^{5,11,18,24,26} Other symptoms include an isolated dry eye in cases in which the geniculate ganglion or geniculate supratrochlear nerve were involved. Since the facial nerve is in close proximity to the vestibulocochlear systems, balance symptoms often occur in association with FNS.

MRI and CT both play a significant role in the evaluation of an FNS. Enhancement along the course of the facial nerve and particularly the geniculate ganglion are highly suggestive of an FNS. Bony erosion around the geniculate ganglion or otic capsule or enlargement of the facial canal is also in keeping with such a lesion. While histological diagnosis remains the gold standard for FNS diagnosis, modern neuroimaging coupled with appropriate clinical history taking and neurological examination can provide reasonable diagnostic accuracy and a basis for determining the next step in the patient's management. In the current study, 36% of patients had a histological diagnosis, and 64% were diagnosed on the basis of radiological and clinical features alone. In the future, high-density diffusion tensor MRI may be of greater value in the evaluation of cerebellopontine angle tumors and the delineation of the neural structures from which they originate as well as those in close proximity.

While conservative management may be pursued in the short term, the absence of treatment often leads to tumor progression and worsening facial nerve function during a moderate to long-term follow up period. Most untreated FNSs grow an average of 1.4 mm per year in the absence of treatment.¹⁹ In a series of 13 patients followed conservatively for a median duration of 6 years (range 1–19 years),

TABLE 4. Summary of the published series of FNSs treated with SRS*

Study	No. of Tumors	Mean/Median Pt Age in Yrs (range)	Pt Sex (M/F)	Mean/Median Tumor Volume in cm ³ (range)	Margin Dose (Gy)	Mean/Median FU Period in Months (range)	Facial Nerve Deterioration	Hearing Preservation	Tumor Control
Herman et al., 2011	2	NA	NA	NA	NA	35	0/2	2/2	2/2
Wilkinson et al., 2011	6	47 (21–65)	2/4	NA†	12.5–13	64	0/6	6/6	4/6
Litre et al., 2007	11	46 (22–87)	4/7	0.89 (4.8–22.5)	10–16	39 (18–84)	0/11	11/11	10/11 (cyst formation)
Madhok et al., 2009	6	40 (19–59)	3/3	1.88 (<1–8.7)	12–12.5	62 (21–85)	0/6	6/6	6/6
Kida et al., 2007	14	45 (28–70)	6/8	5.5 (0.98–20.8)	11–16	31 (12–120)	1/14	13/14	14/14
Current study	42	48 (11–76)	23/19	1.8 (0.04–8.7)	11.0–15	28 (1–139)	4/42	33/42	36/40‡

FU = follow-up; NA = not available; pt = patient.

* Values represent numbers of patients or tumors unless otherwise indicated.

† Tumor diameter was reported as a mean of 16.7 mm.

‡ Tumor control is reported for the 40 patients with reliable radiographic follow-up.

38% had facial nerve functional deterioration.¹⁹ While the timing and choice of approaches remain the subject of considerable debate, treatment of some form is generally required for patients with an FNS.

Resection

One option for management is resection. Microsurgical resection can be undertaken to decompress the nerve,^{1,17,27} debulk or strip the tumor from the nerve,^{12,17,19} or achieve a complete FNS resection with cable grafting or primary anastomosis.^{4,22,25}

In a cohort of FNS patients with reasonable facial nerve function, Wilkinson et al. compared decompression in 21 patients with observation in 15, and they found no significant difference in facial nerve function between the 2 groups after approximately 3 years of follow-up.²⁷ Thus, bony decompression alone does not appear to facilitate preservation of facial nerve function. Tumor resection carries with it the potential for a surgical cure, but it is almost always associated with temporary or permanent facial nerve weakness.

Resection at experienced centers may facilitate preservation of facial function. Symon et al. reported that 9 (90%) of their 10 patients maintained House-Brackmann Grade I or II function after more than 1 year of follow-up.²⁶ In another recent series involving 15 FNS patients with a mean follow-up of 7 years (range 4–11 years), recurrence or regrowth was noted in 26.7% of cases, and the rate of recurrence was markedly higher in patients who had subtotal resection (70%–80%).¹³ Thus, subtotal resection by itself conveys a high chance of recurrence, particularly in the long-term. For patients with favorable preoperative facial nerve function (House-Brackmann Grades I–III), surgical excision with primary anastomosis or grafting rarely affords a level of recovery greater than House-Brackmann Grade IV.¹⁴ These less than optimal neurological results could be partly explained by histopathological findings that have demonstrated nerve fibers running directly through FNSs.²⁵

Radiation Therapy

Fractionated radiotherapy is another option for treat-

ment of FNS. Nishioka et al. reported 4 patients treated with a dose of 50 Gy in 25 fractions and followed for an average of 67.3 months. They showed that 50% of FNSs decreased in size at last follow-up, and facial nerve function was either improved or stable in all 4 patients. In another retrospective study, Hillman et al. (2008) confirmed a high rate of tumor control but a significant hearing loss ipsilateral to the facial schwannoma.⁸ The cochlear division of the eighth cranial nerve and potentially the cochlea itself are radiosensitive structures. A risk of hearing decline of 15% or less has been reported with mean cochlear doses of 45 Gy or lower when delivered in standard fractionation schemes.² While radiation exposure to the cochlea is present in radiosurgery for FNS as well, SRS often can permit greater ability to restrict the radiation dose delivered to the cochlea in accordance with hearing preservation guidelines of 12–14 Gy to the tumor while still potentially delivering an optimal dose to the tumor.²

Stereotactic Radiosurgery

Radiosurgery has proven to be an important tool in the treatment of FNS, with most radiosurgical series demonstrating tumor control rates of 90% or higher for patients with small to moderately sized tumors. Following radiosurgery for an FNS, tumor control is usually achieved, and it is also typically accompanied by neurological preservation or improvement in most patients.

In the published literature, there are only a few radiosurgical series that focus exclusively on FNSs. In a series by Hasegawa et al., 2 patients with FNSs were treated with SRS.⁶ With a follow-up of 42 months, both patients had tumor control and stable facial nerve function.⁶ In another study by the University of Pittsburgh team, Madhok et al. detailed the outcomes of 6 patients treated with radiosurgery and followed for a median of 61.5 months. After delivering a margin dose of 12–12.5 Gy to the tumor in a single session, 3 tumors had regressed, and 3 remained stable. None of the patients had worsening of facial nerve function, and all maintained their preradiosurgical level of hearing.¹⁶ In another study of 11 FNS patients treated using radiosurgery and followed for a median of 39 months, tumor control was achieved in 91% of cases, and no patient demonstrated worsening of facial nerve function.¹⁴ One of

the patients developed a cystic component to the tumor following radiosurgery and required resection of the tumor.

In the current series, tumor control was achieved in 90% of patients at last follow up. Moreover, facial nerve function was preserved in 38 of 42 patients at last follow-up. While these results do not have statistical power or length of follow-up equivalent to that of radiosurgical series of vestibular schwannoma cases, the current study does encompass a period of time over which radiation-induced facial neuropathy should have occurred if it was going to do so. Typically, radiosurgery-induced facial neuropathies occur within 2 years of treatment. In a cohort of 153 vestibular schwannoma patients treated with Gamma Knife radiosurgery, Steiner and colleagues noted facial paresis in 3 patients, and all cases had occurred within 18 months of GKRS.²⁰ Hearing decline, however, may occur in a more delayed fashion after radiosurgery, and the current study may underestimate the rate of hearing decline after GKRS for FNS.³ Nevertheless, extrapolating from the durability of tumor control achieved using radiosurgery to treat vestibular schwannomas, further study of the current study's cohort will likely yield an actuarial control rate greater than 90% at 10 years postradiosurgery.¹⁵ We observed that FNS patients demonstrated a lower risk of temporary or permanent neurological decline when they were treated with a dose of 12.5 Gy or less to the margin of the tumor.

Given the rarity of FNS, Level 1 evidence elucidating the optimal management approach is not likely to be achieved. However, the current study demonstrated more favorable treatment outcomes for FNS patients with a tumor of 1 cm³ or less in volume. We also observed that those patients treated with radiosurgery when they had a House-Brackmann grade of I to III were more likely to maintain this level of facial nerve function at last follow-up. Thus, these findings suggest that early treatment with radiosurgery may be advantageous for patients with FNSs. Radiosurgery for smaller-volume tumors in patients with more favorable facial nerve function led to a greater chance of preservation or improvement in House-Brackmann Grade I–III facial nerve function. Given the reported outcomes with resection and the findings from the current study, this study's demonstration of the benefits of early radiosurgery for small (≤ 1 cm³) FNSs in patients with House-Brackmann grades of I–III is in sharp contrast to the approach taken with microsurgery, where resection is not usually undertaken until there is sufficient mass effect from the tumor and/or facial nerve functional decline to warrant open surgery. Also, early treatment of FNS would be in contrast to the watchful waiting approach taken by some clinicians for vestibular schwannoma patients with intact hearing. Of course, further studies will be required to validate such an approach.

Certainly for those patients with substantial mass effect or for whom substantial diagnostic questions make histological diagnosis prudent, initial resection may remain the treatment of choice. However, when compared with the natural history of an untreated FNS and the alternative treatment options of resection or radiotherapy, the current study findings demonstrate a degree of optimism that radiosurgery offers a favorable benefit to risk profile and may be the preferred approach for those with small to moderate-size tumors.

Study Limitations

The current study represents the largest radiosurgical one to date for patients with FNS. Nevertheless, the study has limitations that merit consideration. Slight variations in dose planning existed between the centers, as would be expected given the retrospective nature of the study. Also, refinement of the Gamma Knife platform (software and hardware) occurred during the study period. Selection bias and the absence of a control arm represent other study limitations. In addition, we do not have data regarding the pre-SRS growth patterns of these tumors. A better understanding of the natural history (both clinical and radiographic) of FNS would be of value to clinicians and the patients that they treat.

Also, not all of the tumors were histologically confirmed to be FNS. Of those tumors that were resected, we did not have available the data on extent of resection. It is possible that some other benign skull base lesions were included in this study. However, any patient with histology inconsistent with a WHO Grade I FNS was excluded from the study. This is true of preradiosurgical histology and histology obtained at the time of resection following failed radiosurgery.

Also, although it has been shown that cochlear dose relates to hearing preservation, we did not routinely limit or collect dose to the cochlea in this series. This may have biased the hearing preservation results in these patients.

Finally, the small statistical power of the study and the modest length of follow-up may have limited the study findings.

Conclusions

Gamma Knife radiosurgery resulted in tumor control and facial nerve preservation in the majority of patients with FNSs. Smaller tumor volumes (1 cm³ or less) and more favorable facial nerve function (House-Brackmann Grade I–III) at the time of radiosurgery portended a more favorable result at last follow-up. While resection continues to be of value for relief of mass effect as well as for histological confirmation in ambiguous cases, radiosurgery used as an early, upfront treatment or an adjuvant one represents a valuable approach to treatment of facial nerve schwannoma patients.

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