GAMMA KNIFE SURGERY FOR FACIAL NERVE SCHWANNOMAS

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OBJECTIVE: Radical resection of facial nerve schwannomas classically implies a high risk of severe facial palsy. Owing to the rarity of facial palsy after gamma knife surgery (GKS) of vestibular schwannomas, functional evaluation after GKS seems rational in this specific group of patients. To our knowledge, no previous similar evaluation exists in the literature.

METHODS: Of 1783 schwannomas of the cerebellopontine angles treated by GKS at Timone University Hospital between July 1992 and May 2003, 11 were diagnosed as originating from the facial nerve. Criteria for this diagnosis were the involvement of the tympanic or mastoid segment of the facial nerve (n = 9) and/or preoperative observation of a facial nerve deficit that had occurred during previous microsurgery (two patients). The rare occurrence of facial palsy after vestibular schwannoma radiosurgery, usually within 18 months of treatment, has been considered only in the patients with more than 2 years of follow-up (n = 9).

RESULTS: Six of these patients experienced a previous spontaneous facial palsy on one (n = 4) or several occasions (n = 2). A normal motor facial function was observed in only three patients before GKS (House-Brackmann Grade II in six patients, Grade III in one patient, Grade IV in one patient). The median follow-up period was 39 months (range, 18–84 mo). At the time of the last follow-up examination, no patients had developed a new facial palsy or experienced deterioration of a preexisting facial palsy and three patients had improvement of a preoperative facial palsy. Ten out of the 11 tumors are stable or decreased in size; in the other, a microsurgical resection of the tumor had been recommended owing to the development of a cyst. Clinical management owing to the specificity and heterogeneity of this group of patients has required the development of an original classification of four anatomic subtypes presenting different clinical and surgical difficulties.

CONCLUSION: This first study demonstrates that radiosurgery allows treatment of these patients while preserving normal motor facial function. Such an advantage should lead to the consideration of GKS as a first treatment option for small- to medium-size facial nerve schwannomas.

KEY WORDS: Facial palsy, Gamma knife surgery, Geniculate ganglion, Radiosurgery, Vestibular schwannoma

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Facial nerve schwannomas (FNS) represent less than 1% of intrapetrous tumors (9), whereas vestibular schwannomas represent 8% (1, 7) of intracranial tumors (10 per million population). Complete tumor resection generally requires the sacrifice of the facial nerve. With successful interposition of a nerve graft or hypoglossofacial anastomosis, a facial nerve recovery is never better than a House-Brackmann Grade III with obvious psychosocial consequences. In this context, any possible therapeutic alternative that could allow better preservation of facial function must be studied. The efficacy of stereotactic radiosurgery has already been demonstrated in the treatment of intracranial benign tumors, especially for vestibular schwannomas. Because facial nerve palsy (10, 15) is an exceptional event after gamma knife surgery (GKS) treatment (<1%), use of GKS seems to be an interesting approach when aiming for functional preservation of the facial nerve.

PATIENTS AND METHODS

A total of 1783 cerebellopontine angle (CPA) schwannomas were operated using GKS and evaluated prospectively between June 1992 and April 2006 at Timone University Hospital. Of these, 11 proved to be FNS. The diagnosis of FNS was based on the following criteria in nine patients: geniculate ganglion invasion and/or invasion of one or more than one segment of the facial nerve in its intrapetrous course and/or temporary spontaneous facial palsy. In the other two patients, the diagnosis was based on previous microsurgical observation. None of these lesions were found in the context of neurofibromatosis. There were seven women and four men with a mean age of 46 years (range, 22–87 yr) at the time of GKS.

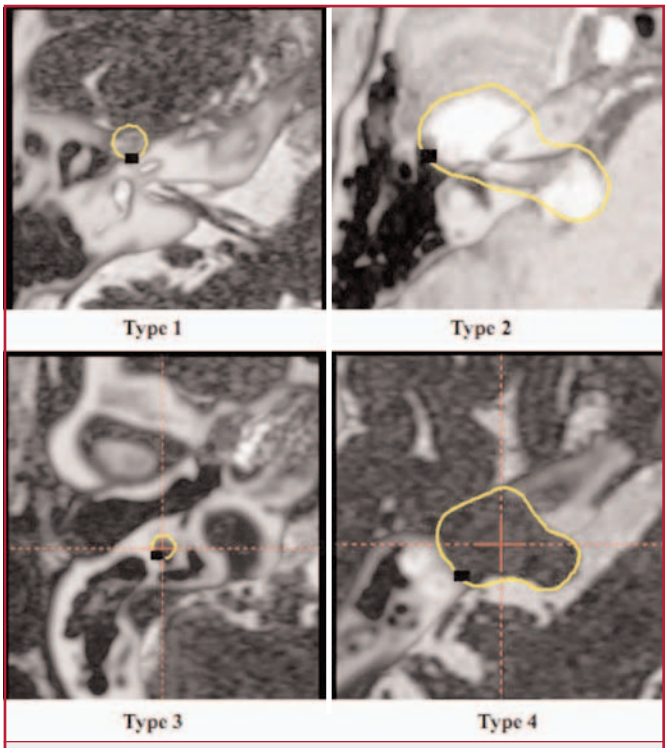

FIGURE 1. *Topographic classification of facial nerve schwannomas. Images are obtained from a fused bone window computed tomographic scan with magnetic resonance imaging (MRI) sequences. The tumor margin is delimited by the* outline, *which represents the 50% isodose.*

Tumor Classification

The lesions have been classified according to an original anatomic classification defined by the topography of the origin of the intrapetrous schwannoma (*Fig. 1*). Functional signs (*Table 1*) were mainly constituted by facial nerve dysfunction in 55% of the patients (facial palsy with one temporary, hemifacial spasm case) followed by cochleovestibular signs in 46% (hypoacousis, tinnitus, vertigo, imbalance) (*Table 2*).

Clinical evaluation of our patients was based on the House-Brackmann facial grading system (5) and the Gardner and Robertson classification (3) for hearing. Facial function was graded before GKS (Grade I, three patients; Grade II, six patients; Grade III, one patient; Grade IV, one patient). The hearing level was graded as Class 1 in three patients, Class 2 in three patients, Class 3 in three patients, and Class 5 in two patients. Another team had previously operated on two patients, but the surgical procedure was interrupted at an early stage to preserve facial function because the tumor was obviously invading the facial nerve. In Patients 3 and 9, the facial nerve was Grade II; the hearing level was Gardner 5 and 3, respectively.

All patients were admitted to the hospital the day before the operation for clinical preparation and completion of all of the required information. Tonal and vocal audiometry, auditory evoked responses, and vestibular tests were systematically performed the day before GKS. The radiosurgical procedure was performed after light sedation. A Type G stereotactic frame was

TABLE 1. Clinical presentation of the patients in this series

Patient no.	Age (yr)/sex	Symptoms	House-Brackmann grade	Hearing grade	Radiological type
1	42/F	Hypoacousis, tinnitus, imbalance, vertigo	Grade I	Class III	Type I
2	66/M	Hypoacousis, tinnitus, imbalance	Grade I	Class III	Type I
3	52/F	Hypoacousis, tinnitus, imbalance	Grade II[a]	Class III	Type IV
4	22/M	Hypoacousis, imbalance, vertigo	Grade II	Class I	Type I
5	46/F	Facial palsy, tinnitus, hemifacial spasm	Grade III	Class I	Type II
6	26/F	Hemifacial spasm, palsy (transient)	Grade II	Class I	Type I
7	87/F	Facial palsy, imbalance, hemifacial spasm, hypoacousis	Grade II	Class V	Type I
8	55/M	Hemifacial spasm	Grade II	Class I	Type I
9	36/F	Hemifacial spasm, tinnitus, hypoacousis, vertigo	Grade II	Class V[a]	Type II
10	48/F	Facial palsy	Grade IV	Class II	Type I
11	25/M	No symptoms	Grade I	Class II	Type I

[a] Postoperative effects.

TABLE 2. Classification of facial nerve schwannomas

Type I	The tumor is localized on the geniculate ganglion.
Type II	The tumor is a dumbbell-shape on the geniculate ganglion, labyrinthine segment, internal auditory canal, and cerebellopontine angle cistern.
Type III	The tumor develops in the tympanic and/or vertical segments of the facial nerve.
Type IV	The tumor develops in the internal auditory canal or the cerebellopontine angle without invasion of the fallopian canal or the geniculate ganglion. This category is difficult to distinguish from the vestibular schwannoma using radiological criteria. In this group, diagnosis was usually based on a previous microsurgical attempt.

positioned under local anesthesia. High resolution magnetic resonance imaging (MRI) scans (Siemens 1.5-Tesla; Siemens Medical Solutions, Erlangen, Germany) were performed in stereotactic conditions. Axial T2 sequence constructive interference in steady state and T1-weighted volumetric MRI pulse sequence scans were performed. Computed tomographic scans in bone windows were systematically performed to identify the petrous bone anatomy and to check the lack of distortion from the MRI scan. The first tumor volume assessment was in volumetric condition, determined by the gamma plan station. The mean tumor volume was 888 mm^3 (range, 48–2253 mm^3). The mean peripheral isodose was the 50% isodose with an average isodose of 13 Gy (range, 10–16 Gy) (*Fig. 2*). The average number of isocenters was 13 (range, 1–33).

The final decision in terms of the marginal dose to be used takes into account various different data, including the results of functional testing (hearing and facial levels) and the topography of the FNS. After removal of the frame, patients spent one night in the neurosurgical department. After clinical evaluation the following day, all were able to return to their normal daily and professional activities.

RESULTS

The patients were sequentially followed at 6 months and 1, 2, 3, 5, and 7 years. Each control included clinical examination and MRI scans. The mean clinical and radiological follow-up period was 39 months (range, 18–84 mo). Nine patients have been followed for more than 2 years (*Table 3*).

Tumor Control

At the 6-month follow-up examination, tumor size was considered unchanged in six patients and increased in five; at the 1-year follow-up examination, tumor size decreased in three of these. For the nine patients who were followed for more than 2 years after treatment, criteria of decreased size was obtained in three patients, stabilized in five patients, and increased in one patient. In this latter case, we observed a global increase of tumor volume owing to cyst development (*Fig. 3*). Microsurgical resection of this tumor was recommended.

Functional Clinical Result

We observed an improvement of the functional signs in 11 patients. Resolution of hemifacial spasm was observed in two patients. In one patient, a total facial palsy recovered. An improvement of facial nerve function from Grade II to Grade I was observed in two patients, and improvement of facial nerve function from Grade 4 to Grade 2 was observed in one patient. Facial grade remained unchanged in the other patients (*Table 4*).

One patient experienced recovery of balance problems and a clear decrease in tinnitus, which led to hearing improvement. Patients with a useful hearing level at the preliminary

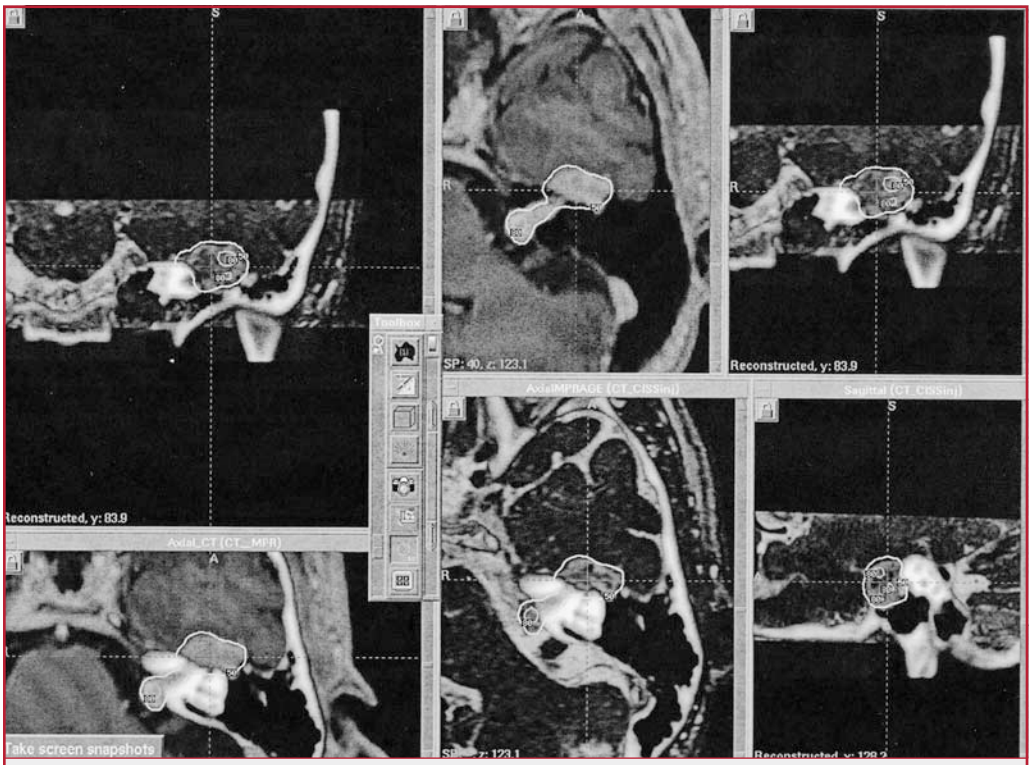

FIGURE 2. *Dose planning for a Type 2 FNS.*

TABLE 3. Treatment and follow-up of our patients

Patient no.	Dose (Gy)/ no. of shots	Initial tumor volume (mm^3)	Volume at 6 months	Loss of contrast enhancement	Follow-up (mo)/ volume of tumor
1	10/18	1672	Stable	N	54/decreased
2	12/14	963	Stable	Y	24/stable
3[a]	12/33	2253	Stable	N	28/stable
4	12/19	644	Increased	Y	48/decreased
5	12/7	385	Stable	N	18/stable
6	16/1	48	Increased	N	84/decreased
7	11/17	207	Stable	Y	36/stable
8	12/1	87	Stable	N	30/decreased
9[a]	10/12	1669	Increased	Y	60/increased
					(cyst)
10			Increased	N	20/stable
11			Stable	N	32/stable

[a] Subtotal surgical removal.

examination maintained this at time of their last follow-up examination. No patients improved from non-useful to useful hearing after radiosurgery.

DISCUSSION

The Facial Nerve

The facial nerve emerges from the pontomedullary sulcus near the cochleovestibular nerve, courses in the CPA transversally, and enters the internal auditory canal with an anterosuperior position. At the fundus of the internal auditory canal, the facial nerve penetrates the facial canal just in front of a small osseous vertical crest known as the "Bill bar." The first horizontal portion of the facial canal contains the facial nerve (labyrinthine segment) and the intermediate nerve of Wrisberg. The facial canal quickly leaves the region of the vestibule and approaches the geniculate fossa, which contains the geniculate ganglion. It then courses posteriorly and slightly laterally, thus forming an acute 75-degree angle with variations that have been extensively described in textbooks (13). It extends from the geniculate fossa to the posterior wall of the tympanum and displays a close relationship with the loop of the lateral semicircular canal. The vertical portion of the facial canal extends from the second turn to the stylomastoid foramen. It is nearly rectilinear and forms an angle ranging from 95 to 125 degrees with the tympanic portion of the facial canal. It runs through the mastoid process, and thus merits the name of mastoid portion.

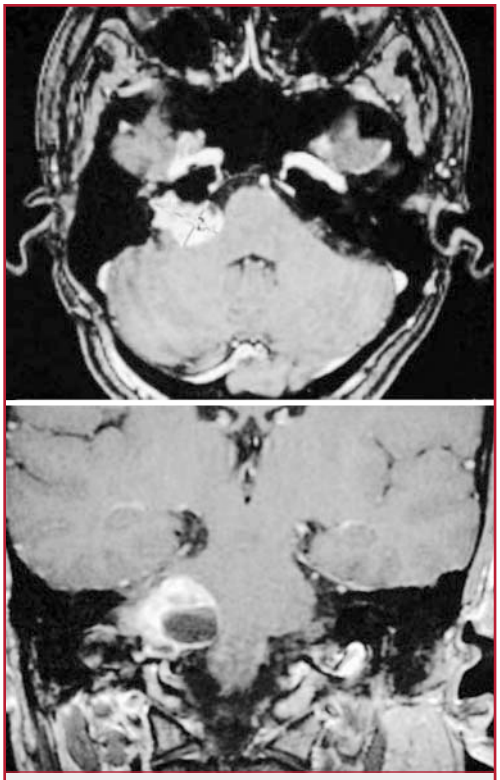

FIGURE 3. *MRI scans showing the evolution of an FNS after radiosurgery, with development of a cyst.*

Diagnosis of FNS

FNS are rare tumors affecting both men and women equally. Such tumors may be diagnosed regardless of the age of the patient, but middle-aged people are predominantly affected. Clinical manifestations are heterogeneous, depending on tumor origin and extension. The primary symptoms are linked to facial nerve dysfunction, regardless of the location of the tumor on the facial nerve. Hemifacial weakness or hemifacial spasm of sudden or progressive onset is the major symptom. Transient and recurrent deficit is typical of the disease. Isolated dry eye or dysgeusia may occur in cases of isolated involvement of the geniculate ganglion or geniculate suprapetrous nerve.

TABLE 4. Hearing status of patients with initial useful hearing[a]

Patient no.	Type	Volume (mm^3)	Dose Gy	PTA before (dB)	GR before	Follow-up (yr)	PTA after (dB)	GR after	Delta (dB)	Functional deficit (GR)
6	I	48	16	5.00	1	5	13.75	1	8.75	No
4	I	644	12	16.25	1	3	43.75	2	27.50	No
8	I	87	12	25.00	1	3	27.50	1	2.50	No

[a] PTA, pure tone average; GR, Gardner Robertson score.

TABLE 5. Comparison of series from the literature[a]

Series (ref. no.)	No. of patients	Therapy	Evolution		Follow-up, recurrence/ tumor progression
			Hearing	Facial function	
Sherman et al., 2002 (17)	10	Microsurgery	2 worsened, 7 stable, 1 enhanced	2 worsened, 5 stable, 3 enhanced	39.8 mo, 1 patient
Fichten et al., 2006 (2)	7	Microsurgery	6 worsened, 1 enhanced	7 worsened	
Hasegawa et al., 1999 (4)	2	Radiosurgery Gamma knife radiosurgery	1 enhanced, 1 stable	2 stable	42 mo, 0
Mabanta et al., 1999 (11)	2 with 6 schwannomas	Radiosurgery LINAC	2 stable	2 stable	32 mo, 0
Ulku et al., 2004 (18)	4	Surgery	Not available	1 stable, 1 worsened, 2 enhanced	38 mo, 0
Current series	11 (2 with surgery)	Gamma knife radiosurgery	11 stable	3 enhanced, 8 stable	39 mo, 1 patient[b]

[a] LINAC, linear accelerator.
[b] Patient already operated (evolution with a cyst).

Otological and vestibular manifestations are also frequently encountered. Imbalance and vertigo indicate that the tumor affects the vestibular system in the case of CPA or internal auditory canal involvement. Sensorial hearing deficit or tinnitus indicates a cochlear nerve dysfunction in the CPA or internal auditory canal, although the cochlea is generally not directly involved by the tumor. Conductive hearing loss or the sensation of a blocked ear is typical of middle ear invasion. Diagnosis is based on high-resolution neuroimaging. Computed tomographic scans show an enlargement of a segment of the fallopian canal with homogeneous bone erosion, whereas MRI scans show the enhanced tumor mass on a segment of the nerve. In cases of geniculate fossa invasion, the tumor may extend upward in the middle fossa, and may sometimes display cystic transformation. In an extensive review of the literature conducted over more than three decades (17), the tumor location was described as follows: labyrinthine, 43%; tympanic, 42%; vertical segment, 36%; and CPA, 17%. Owing to refinement in imaging techniques, the diagnosis is now made earlier and the origin of the tumor can be more accurately defined. It has recently been postulated that FNS predominantly develop from the geniculate ganglion and petrosal nerves (19). Taken collectively, the data obtained from the range of clinical presentation and neuroimaging findings led us to establish an original classification of FNS.

Timing and Modalities of Treatment

FNS are benign tumors that are supposed to display slow growth, but little is known about their natural history. In a recent study, a cohort of 13 patients was followed conservatively (12). It has been shown that facial nerve deterioration was observed in 38% of patients during a median follow-up period of 6 years (range, 1–19 yr). This information is important to take into account during the treatment decision process. When treatment is recommended, microsurgical radical removal was traditionally considered the treatment of choice. Technical aspects of tumor removal depend on tumor origin, extension, hearing level, and preexisting deficits. The main consequence of tumor extirpation is facial nerve palsy. The realization of facial nerve anastomosis using an interposed cable graft or hypoglossofacial anastomosis partially reduces the deficit. However, even in the best case scenario, the level of recovery that can be reached is never better than Grade III. In rare cases of extrafascicular growth of the nerve or in the case of geniculate suprapetrous nerve involvement, the nerve may be preserved (17), but this situation cannot be planned before the operative period. Consequently, several authors propose a "wait and see" strategy and recommend radical resection after deterioration of facial motor nerve function. In parallel, it has been shown that the occurrence of facial nerve palsy never exceeds 0.5% after GKS treatment (14) with a 97% rate of long-term tumor control, whereas the facial nerve is always in close relationship with the tumor that is exposed to radiation. Taken collectively, these observations led us to consider that radiosurgery could represent a safe and efficient alternative to conventional microsurgical removal of FNS. The present study is the first report that focuses on a series of patients who have been treated homogeneously with a state of the art technique. In 1999, Mabanta et al. (11) reported the absence of short-term worsening of facial motor function in two patients treated with linear accelerator-based radiosurgery, but no definitive conclusion could be drawn from these reports (*Table 5*). In terms of tumor control, the length of the follow-up period is still too short to provide definitive data concerning tumor control. However, 50% of the patients have been followed for more than 3 years without tumor growth. Notwithstanding, this period is long enough to provide reliable functional results. Facial nerve function did not worsen for any patient, which compares favorably

to microsurgery. This result is also interesting because most patients presented with facial nerve deficit before radiosurgery; thus, it would have been expected that radio-induced neuropathy might occur more often when the nerves have already been damaged. This situation is comparable to what has already been observed for meningiomas affecting oculomotor nerves. In these cases, it is usual to observe a recovery of the oculomotor palsy after GKS (16). Of particular interest is the question of whether or not complete palsy may recover after GKS, because restorative microsurgery of the nerve is able to improve the motion to Grade III. Hearing level was also preserved in all patients in the present series, whereas the intrapetrous neuro-otological structures are jeopardized by open microsurgery.

CONCLUSION

Surgical management of FNS yields the risk of severe and permanent deficit of cochleovestibular and facial nerve deficit. In the present study, we have shown that GKS treatment was an interesting option, permitting tumor control and functional preservation in small to mid-sized tumors. Treatment of additional patients with longer follow-up periods after GKS should confirm these preliminary data. The decision regarding early proactive treatment remains debatable. If tumor volume exceeds a 3-cm diameter or if the facial nerve deficit is severe and permanent, it may be more valuable to recommend microsurgical removal and nerve reconstruction.

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COMMENTS

Virtually no center outside of Marseille has achieved such remarkable results in managing acoustic neuromas or the much-rarer facial schwannomas. This report describes the outcomes, which are generally quite favorable, of patients who received radiosurgery as a primary management strategy. Most patients were diagnosed by characteristic imaging, although two had prior surgery. The major outcome factor after microsurgery was a virtually certain facial palsy. After gamma knife surgery, functional facial nerve preservation as well as hearing preservation were possible. In general, schwannomas seemed to respond well, although each had its own idiosyncrasies. In our experience, trigeminal nerve sheath tumors are much more likely to have transient tumoral enlargement at 4–12 months, and this is sometimes associated with temporary sensory dysfunction. In contrast, transient enlargement of acoustic neuromas occurs in fewer than 5% of patients. Facial schwannomas and jugular bulb schwannomas also respond well to radiosurgery, and in most cases this includes excellent motor preservation. This report argues favorably for the role of gamma knife surgery as a primary management strategy: patients can expect tumor control, hearing preservation (in those who had hearing at presentation), and facial nerve function maintenance.

L. Dade Lunsford
Pittsburgh, Pennsylvania

Litre et al. from the Timone University Hospital in Marseille have documented, for the first time, outcomes after radiosurgery of facial nerve schwannomas. The logic of this approach is clear: facial nerve outcomes after resection are typically poor, whereas the chance of a facial nerve deficit after vestibular schwannoma radiosurgery is exceedingly low. Using a dosing regimen based on their vast experience with vestibular schwannoma radiosurgery (median tumor margin dose, 13 Gy), 10 out of 11 tumors have remained stable or decreased in size. Four patients had improved facial nerve function, and three patients with Gardner-Robertson Class 1 hearing before radiosurgery retained either Class 1 (n = 2 patients) or Class 2 (n = 1 patient) hear-

ing at their last follow-up examination. Although these are clearly early results and more follow-up reporting is needed, the decision to manage patients with facial nerve schwannomas using radiosurgery as opposed to surgical resection makes perfect sense if one considers the risk-to-benefit ratio.

Bruce E. Pollock
Rochester, Minnesota

The authors report a small series of 11 patients with facial schwannomas who were treated using stereotactic radiosurgery. They have shown that tumor control rates were high (91%) with no worsening in preradiosurgery treatment function. Also interesting is that 4 of the 11 patients (36%) had improvement in their facial nerve function of one or two levels on the House-Brackmann grading scheme. The pretreatment hearing level was preserved in all patients who had functional hearing before their radiosurgery; this finding is significant because hearing loss is a risk associated with open surgery for these tumors. Another finding, which is similar to observations of acoustic neuroma patients, is that there can be a transient increase in tumor size during the first 12 months as the tumor undergoes necrosis from treatment. Although additional follow-up is needed for these patients, it appears that radiosurgery is a viable treatment option for facial schwannomas, as the procedure appears to convey a high rate of tumor control with low risk to hearing and facial nerve function.

Steven D. Chang
Stanford, California

Litre et al. present detailed information on 11 presumed facial nerve schwannomas that were followed-up for at least 18 months after radiosurgical treatment. No patients experienced worsening of facial function and three patients' symptoms improved. One patient experienced enlargement of a cystic tumor; the remainder of the tumors were controlled. This report reflects the largest series of facial nerve schwannomas treated with radiosurgery and represents a significant contribution to the literature.

A cautionary note: not every enhancing lesion that tracks along the facial nerve and causes facial paralysis is a facial schwannoma. Other pathological entities, including hemangioma and simple inflammation can have a very similar appearance on magnetic resonance images. Radiosurgery may indeed become a preferred treatment for facial schwannomas, given the morbidity of surgery, but longer follow-up periods of larger numbers of patients are needed.

William A. Friedman
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In this article, Litre et al. report on their experience with gamma knife surgery for facial nerve schwannomas. As compared with their eighth nerve counterparts, these tumors are relatively rare. In a busy center such as Timone, these tumors comprise fewer than 1% of all cerebellopontine angle tumors that were treated.

It is not surprising that 10 out of 11 patients demonstrated no tumor growth on their last follow-up neurological imaging. It has been shown with vestibular and trigeminal nerve schwannomas that gamma knife surgery affords a high rate of local tumor control. The significant finding in this study is that no patients experienced new or worsening facial palsy, and that three patients actually had improvements in preoperative facial weakness.

Our results at the University of Virginia for non-eighth nerve schwannomas treated using the gamma knife have been similarly encouraging. A dose of 11–15 Gy to the periphery typically effectuates local tumor control and maintenance or improvement of neurological function. We have found magnetization-prepared rapid-gradient echo and constructive interference in steady-state magnetic resonance sequences very useful for dose planning. In seventh or eighth nerve schwannomas, limiting the cochlear and intracanalicular doses whenever possible may prove efficacious in terms of auditory function preservation.

Such results certainly demand longer term follow-up. A larger patient population would be preferable, but, given the relative rarity of these tumors, single-institution experiences will be fairly limited. Clearly, microsurgery should not be excluded; in the case where diagnosis is uncertain or the tumor is large and exerting mass effect, extirpation should be performed. However, the intermediate results from this facial nerve schwannoma series suggest that gamma knife surgery may represent the treatment of choice for small to moderately sized tumors.

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