Multimodality Management of Trigeminal Schwannomas

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Abstract

Patients presenting with trigeminal schwannomas require multimodality management by a skull base surgical team that can offer expertise in both transcranial and transnasal approaches as well as radiosurgical and microsurgical strategies. Improvement in neurologic symptoms, preservation of cranial nerve function, and control of mass effect are the primary goals of management for trigeminal schwannomas. Complete surgical resection is the treatment of choice but may not be possible in all cases. Radiosurgery is an option as primary management for small- to moderate-sized tumors and can be used for postoperative residuals or recurrences. Planned surgical resection followed by SRS for residual tumor is an effective option for larger trigeminal schwannomas. The endoscopic resection is an excellent approach for patients with an extradural tumor or tumors isolated to the Meckel cave. A detailed analysis of a tumor and its surroundings based on high-quality imaging can help better estimate the expected outcome from each treatment. An expert skull base team should be able to provide precise counseling for each patient's situation for selecting the best option.

Keywords

- ▶ trigeminal schwannomas
- ▶ radiosurgery
- ▶ skull base
- ▶ transnasal approaches
- ▶ endoscopic resection

Introduction

Trigeminal schwannomas are the most common among nonvestibular schwannomas and account for 1 to 8% of intracranial schwannomas and 0.1 to 0.5% of intracranial tumors. The peak incidence is during the third and fourth decades but can occur at any age including young children. Trigeminal schwannomas can arise anywhere between the root and the distal extracranial branches of the trigeminal nerve.¹ Their clinical presentation depends on tumor location and size. The most common presenting symptoms include facial hypoesthesia, headache, dizziness, and ataxia.

Imaging and Tumor Classification

Careful evaluation of high-quality preoperative magnetic resonance imaging (MRI) often establishes the type of trigeminal schwannoma, approximates tumor volume, and defines its relationship with neighboring neural and vascular structures. These parameters can help guide the management strategy. In cases of large tumors computed tomographic (CT) scans could show enlargement of superior orbital fissure, the foramen ovale, or foramen rotundum, as well as erosion of the middle fossa and the petrous apex. Preoperative angiography is usually not needed as trigeminal schwannomas are usually moderately vascular.

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The tumors can be categorized into the four types according to Day and Fukushima's modified version² of the Jefferson classification system. Jefferson categorized the tumors into the following three types³: type A, tumor of the gasserian ganglion in the middle cranial fossa; type B, tumor of the roots of the trigeminal nerve in the posterior fossa; and type C, tumor occupying both the middle fossa and the posterior fossa. Day and Fukushima² added a fourth category, type D, for tumors with an extracranial extension that arises from the branches of the trigeminal nerve. The tumors can be further subclassified depending on nature, location, and relationship to the brainstem.⁴ A more recent classification into six different types has been proposed.⁵

- Type P: posterior fossa tumor in the subdural space
- Type M: middle fossa tumor in the interdural space
- Type E: extracranial tumor in the epidural space
 - E1: tumor in the orbit
 - E2: tumor in the pterygopalatine and infratemporal fossae
- Type MP: dumbbell-shaped tumor in the middle and posterior fossae
- Type ME: dumbbell-shaped tumor in the middle fossa and extracranial space
- Type MPE: tumor involving the posterior fossa, middle fossa, and extracranial space

Chen et al noted 62% tumors in the middle fossa or both the middle fossa and the posterior fossa.⁶ Goel reported 81% tumors occupying the middle fossa and the posterior fossa.⁷ Wanibuchi reported 66% tumors occupying the middle fossa and the posterior fossa. The trigeminal schwannomas usually displace the neighboring neural and vascular structures, as opposed to the invasion of the venous spaces of the cavernous sinus or encasement of the cavernous sinus portion of the internal carotid artery (ICA).⁸

Management

The scientific literature on trigeminal schwannomas is strikingly devoid of publications discussing the natural history of these lesions. Contrary to vestibular schwannomas that can present with hearing loss, imbalance, or tinnitus even at a small size, small trigeminal tumors rarely cause symptoms and are often incidental findings on MRIs obtained for other reasons. Given the benign histopathologic features of schwannomas, it behooves us to extrapolate from the vestibular schwannoma data and consider incidentally found trigeminal tumors as likely slow-growing or possibly tumors not growing at all. Conservative management with serial imaging should therefore be considered and discussed in incidentally found trigeminal schwannomas. The correlate of this position is that any small tumor of the trigeminal nerve presenting with rapidly progressing neurologic symptoms should bring into questions the diagnosis of benign schwannoma and should either be followed at very close interval or be considered for surgical biopsy or resection to rule out other more aggressive lesions.

The treatment goals for trigeminal schwannomas are heavily dependent on the tumor presentation. Even if com-

plete resolution of facial numbness is rarely achieved by either surgical resection or stereotactic radiation, a numb face can be considered an excellent outcome in a patient whose face was numb at presentation and who was suffering from symptoms of brainstem compression. In contrast, even a partially numb face, in one of the divisions of the trigeminal nerve, can constitute a lifelong handicap (corneal anesthesia, cheek biting, or drooling) in a patient who was asymptomatic and fully functional prior to treatment.

As is often the case in the management of potentially benign lesions involving structures whose damage has a high impact on a patient's quality of life, management decisions have to be individualized to each tumor, each patient, and each community to minimize morbidity and maximize safety.

Observation or Biopsy

Owing to the lack of scientific reports supporting or questioning the value of observation, the authors will just comment on the concept, discussed previously, that observation should always be considered in asymptomatic small tumors. The basis of a watch-and-wait strategy should be a near-perfect concordance between imaging characteristics, clinical presentation, and patient's medical and social factors that would make the incidentally discovered tumor highly likely to be a benign schwannoma in a patient able to undergo regular follow-up.

In a study of growth rate O'Reilly et al found that five out of nine nonvestibular cranial schwannomas (NVs) grew significantly with a growth rate of greater than 5% per year during a mean period of 35 months. These authors noted that the proportion of patients exhibiting growth and the rate of growth were higher compared with unilateral sporadic vestibular schwannomas, but lower than those with neurofibromatosis type 2 (NF2). These authors suggested early active treatment for patients with symptomatic progression.⁹ Fisher et al in a study of prevalence and location of cranial nerve schwannomas found that NVS occurred in approximately half of NF2 sample with trigeminal nerve being the most common site. These authors also noted that, in general, NVs were slow growing, in contrast to vestibular schwannomas, and recommended monitoring these tumors until documented growth.¹⁰

If the radiographic characteristics are not typical for a schwannoma, the clinical course suggests a more rapidly progressing tumor, or the patient's comorbidities question the diagnosis of trigeminal schwannoma, then serious consideration should be given to obtaining tissue for histopathologic analysis. This will most often mean taking the patient to surgery for complete or partial tumor resection, but in cases where less invasive options are considered more appropriate, a simple biopsy of Meckel cave can be achieved via at least two minimally invasive routes.

A percutaneous biopsy^{11,12} of Meckel cave can be obtained by the same approach as in the percutaneous treatment of trigeminal neuralgia (balloon compression, radiofrequency, or glycerol rhizotomy). The entry point is 2.5 cm lateral to the labial commissure, and using fluoroscopic guidance, the needle is advanced through the cheek, medial to the ramus

of the mandible, all the way to the inferior skull base where the foramen ovale is penetrated. The importance of fluoroscopic guidance cannot be underestimated to avoid penetrating, the inferior orbital fissure, or the jugular foramen.¹³ This technique is suitable for all tumors that involve Meckel cave and/or the third division of the trigeminal nerve.

A biopsy of Meckel cave can also be obtained by an expanded endonasal approach. Compared with the percutaneous route, it is a more time-consuming procedure but allows a wider exposure and therefore a greater access to the lesion. All the divisions of the trigeminal nerve can be exposed endoscopically and the expanded endonasal approach can be converted from a simple biopsy to a tumor resection if necessary.

Surgical Resection

Both open and endoscopic techniques are currently used to resect trigeminal schwannomas and both strategies involve different steps in a “building blocks” strategy to gain the degree of exposure dictated by the tumor. The choice of the strategy (endoscopic vs. open) is still a matter of debate but might one day be dictated by the tumor-nerve anatomical situation. Currently, no reliable way of determining this relationship is available, but rapid progress in acquisition and processing of diffusion tensor imaging MRI sequences will soon allow to determine preoperatively whether the trigeminal nerve fibers are medial or lateral to the tumor. Transcranial approaches might then appear more suited for tumors that displace the trigeminal nerve medially while endonasal techniques might be more suited for tumors displacing the nerve laterally.

Open and Endoscopic Assisted Approaches

The different classifications of trigeminal schwannomas, discussed earlier, directly impact the choice of surgical approach used to resect these tumors. The trigeminal nerve is simultaneously located in the intradural subarachnoid space of the posterior fossa, interdural subarachnoid space of Meckel cave, interdural space of the lateral wall of the cavernous sinus, and finally extradural space of the extracranial portions of V1–V2 and V3. The exact location of the tumor along these different segments of nerve dictates a specific “building block” for a transcranial approach. Transitional areas in tumors that are mainly in one compartment with a minor portion in a different compartment generate, as usual, the greatest interest and discussion. Tumors that are exclusively located in the posterior fossa can be safely removed using a retrosigmoid craniotomy. Whether in the semi-sitting or park bench position, the resection can usually be accomplished with minimal to no retraction of the cerebellum. Some critics of this approach contend that the operative field is quite deep and that the facial and vestibulocochlear nerves are put at risk because of their more superficial location than the trigeminal nerve in a retrosigmoid approach to the cerebellopontine angle. If this is a correct statement in the anatomical specimen, the surgical view is often more forgiving, since medium to large trigemi-

nal schwannomas push the VII–VIII cranial nerves complex inferiorly and therefore significantly increase the size of the working window. Furthermore, as the root of the trigeminal nerve is located on the lateral surface of the pons, these tumors often push the brainstem anteriorly and medially, thereby “opening” the window from a retrosigmoid craniotomy, as opposed to petroclival meningiomas that can sometimes have a “cap” of brainstem on their dorsolateral surface that “closes” the access from a retrosigmoid approach. The same working area (lateral surface of the pons and a trigeminal schwannoma located on the cisternal segment of the nerve) can also be safely accessed through a middle fossa approach and drilling of Kawase’s triangle of the petrous apex. The advantages of a lateral approach include a shorter working distance and the ability to work on the anterolateral surface of the brainstem with no cerebellar or brainstem retraction. The Kawase approach to trigeminal schwannomas has, nevertheless, some disadvantages such as a longer exposure time and the need to apply retraction on the temporal lobe. The temporal lobe retraction that can significantly impact the outcome is not so much the extradural retraction applied during the drilling phase of the approach, but rather the intradural retraction applied directly on the temporal lobe during the tumor resection. If the tumor resection is expected to last several hours, one has to anticipate some degree of temporal lobe damage whose clinical significance is heavily dependent on the side of the approach and the laterality of speech. Use of a lumbar drain and osmotic diuresis to obtain optimal brain relaxation help diminish the amount of retraction on the temporal lobe but do not eliminate it completely. The retrosigmoid and Kawase approach can be combined in what has been called the *retrosigmoid intradural suprameatal approach*. In this approach, a standard retrosigmoid craniotomy is combined with an intradural suprameatal drilling of the petrous apex to reach the middle fossa extension of a trigeminal schwannoma with or without endoscopic assistance.⁴ Alternatively, a transpetrosal approach using a posterior petrosectomy and partial labyrinthectomy can also be used. In cases with marked expansion of Meckel cave and a dominant middle fossa component, a purely anterior approach becomes a viable option via either a pterional or FTOZ (frontotemporal-orbitozygomatic) craniotomy utilizing an extradural dissection. In this situation, the expanded Meckel cave becomes the corridor by which the posterior fossa component is resected.

For tumors extending more anteriorly in the middle fossa and more inferiorly in the infratemporal fossa, a zygomatic osteotomy can be added to the subtemporal approach and partial resection of the middle fossa floor can expose most of the inferior extent of trigeminal schwannoma. Dedicated infratemporal fossa approaches such as described by Fish are rarely, if ever, needed in the resection of trigeminal schwannomas. The additional time and effort required for any of the extensive skull base approaches described earlier needs to be put into context that these are benign tumors in which a subtotal resection and postoperative radiosurgery is often a very reasonable strategy.

Purely Endoscopic Approaches

Because of its paramedian and parasellar location, Meckel cave is amenable to an endoscopic transnasal approach. Although most published case series on the endoscopic resection of trigeminal schwannomas are small, their results are promising¹⁴ and signal a possible paradigm shift in the management of these tumors. The clear advantage of endoscopic approaches is the absence of retraction on either the temporal lobe, the cerebellum, or the brainstem. The disadvantages of the endoscopic route include the increased incidence of cerebrospinal fluid (CSF) leak and the location of the carotid artery in the middle of the surgical field, between the tumor and the surgeon. Careful analysis of the carotid artery and how it is displaced by the tumor plays a major role in the feasibility and planning of an endoscopic approach to Meckel cave tumors. The gasserian ganglion is located just posterior and slightly medially to the lacerum segment of the ICA, the V2 and V3 branches lie laterally to the ascending segment (sometimes called *paraclival segment*) and the horizontal segment of the cavernous ICA, and finally the V1 branch lies mostly superior to the horizontal segment of the cavernous ICA. A trigeminal schwannoma can therefore displace the carotid in virtually any direction, depending on its size and its exact anatomical epicenter, thereby opening up specific corridors and closing others, during an expanded transnasal approach. In-depth knowledge of the anatomical landmarks to the lacerum, paraclival, and cavernous segments of the ICA (in particular the vidian nerve) and the use of a micro Doppler to positively identify the ICA prior to incising the dura are part of the surgical strategy that allows for a safe and successful resection of a trigeminal schwannoma from an expanded transnasal route. The part of the tumor extending into the posterior fossa (the retrocarotid space when coming from a transnasal approach) will require drilling of the posterior clinoid process and identification of the abducens nerve in Dorello's canal by means of a stimulating probe. The ideal targeted endoscopic approach would involve an extended sphenoid sinus approach or a pterygomaxillary approach with the vidian nerve and the carotid artery as anatomical identifiers to the location of the Meckel cave.

Surgical Results

Outcome from surgical resection has been reported in several series, and although some authors have reported improvement in preoperative hypesthesia^{1,7,8} in most cases, the presenting symptoms persisted or worsened after surgical resection. Goel et al⁷ reported improvement in the preoperative trigeminal sensory deficit in 40% of the patients but worsening in 27%. Wanibuchi et al⁸ noted improvement in 16%, no change in 73%, worsening in 12%, and new hypesthesia in 22% of patients. Chen et al reported unchanged facial hypesthesia in 72% and improved in 28%. Other frequent symptoms include those related to involvement of adjacent cranial nerves in the cavernous sinus.^{1,7,8} Wanibuchi et al⁸ reported diplopia in 20% (86% due to deficit of the abducens nerve and 14% due to deficit of the oculomotor nerve). Al-Mefty et al¹⁵ noted diplopia in 52% (40% due to sixth nerve deficit). Chen et al reported diplopia in 18% of patients (80%

due to abducens nerve deficit and 20% due to oculomotor nerve). The diplopia improved in 70% of patients postoperatively. In a recent study of 20 patients, Samii et al reported improvement in facial pain in all patients and cerebellar ataxia in three of four patients.⁴ Jeong et al noted improved or unchanged trigeminal symptoms in 51% but worsening of facial hypesthesia in most of the cases.¹⁶ Fukaya et al studied 57 patients who had surgical resection for trigeminal schwannoma. While 42 of 45 patients (93%) who had skull base surgery achieved a complete tumor resection, 39 of 57 patients (68%) developed complications that included cranial neuropathies, brain contusion, or CSF leakage.¹⁷ Sharma et al evaluated 68 patients with trigeminal schwannomas who had surgery.¹⁸ These authors reported 76% rate of complete tumor resection, 2% mortality, and 15% permanent morbidity (►Table 1).

Stereotactic Radiosurgery

Although advances in microsurgical techniques have significantly decreased the perioperative morbidity of trigeminal schwannoma surgery, the potential for associated complications such as cranial neuropathy and CSF leak continue to present challenges to complete tumor resection. These tumors often progress after subtotal resection.²³ Stereotactic radiosurgery (SRS) is a reasonable alternative to re-resection for such patients given the risks of reoperation. It has been used as either a primary or an adjunct treatment for trigeminal schwannomas. Radiosurgery offers the advantages of a high tumor control rate and a low rate of radiation associated complications.

Several reports have highlighted the role of radiosurgery in the multimodality management of these tumors. The results of recent radiosurgical series are shown in ►Table 2. Tumor control rates ranged from 79 to 93% with a mean follow-up period of 4 to 8 years for patients with a mean tumor volume of 4.2 to 8.7 mL. SRS is associated with a very low risk of new neurologic deficit. Most radiosurgical series report few adverse radiation effects other than facial hypoesthesia or facial pain which are related to the fifth cranial nerve. Hasegawa et al in a recent study showed that morbidities caused by adverse radiation effects were limited to facial hypoesthesia or facial pain 10% of patients. In contrast to surgical resection, injuries to the brain, vascular, or cranial nerve other than to the fifth cranial nerve are unlikely after gamma knife surgery (GKS). On the contrary, 11 to 70% of patients experienced some degree of improvement of preexisting signs or symptoms.

Pollock et al reported outcomes after GK SRS in 10 patients with trigeminal schwannoma and reported tumor control in 9 patients. New or worsening trigeminal dysfunction developed after GKS in three patients. Nettel et al reported that tumor shrinkage or no new growth was observed in 20 (91%) of 22 patients harboring trigeminal schwannomas at a median imaging follow-up of 40 months. Radiosurgery was performed using a median marginal dose of 15 Gy (range 13–20).²⁵ These authors demonstrated that 52% of patients had an improvement in symptoms and that two patients

Table 1 Trigeminal schwannoma: surgical series

Author	Year	Tumor classification and No. of cases					Radical removal (%)	Mortality (%)	Morbidity (%)
		A	B	C	D	Total			
McCormick et al ¹⁹	1988	6	5	2	1	14	6 (43%)	0	78
Pollack et al ²⁰	1989	6	4	5	1	16	12 (75%)	0	6
Dolenc ²¹	1994	NA	NA	NA	NA	40	40 (100%)	0	25
Konovalov et al ²²	1996	42	26	30	13	111	86 (77%)	3	87
Yoshida and Kawase ⁵	1999	4	5	10	8	27	20 (74%)	0	74
Goel et al ⁷	2003	29	7	30	7	73	51 (70%)	3	7
Pamir et al ¹	2007	5	2	9	2	18	17 (94%)	0	28
Wanibuchi et al ⁸	2012	39	22	33	14	105	86 (82%)	0	9
Chen et al ⁶	2014	13	10	21	11	55	52 (95%)	0	5
Fukaya et al ¹⁷	2010	15	12	26	4	57	46 (81%)	2	68
Samii et al ⁴	2014	8	1	8	4	20	15 (75%)	0	4
Jeong et al ¹⁶	2014	20	20	9	0	49	47 (95.9%)	0	18

experienced newly developed symptoms, including transient facial weakness and worsening of preradiosurgery facial numbness. Huang et al described outcomes for GK SRS in 16 trigeminal schwannoma patients²⁴ and reported 100% tumor control and no complications at a mean follow-up period of 44 months. Mabanta et al described the outcomes of linear accelerator SRS in seven patients with trigeminal schwannoma.³⁴ After an average follow-up period of 32 months, the tumor control rate was 100%. Phi et al treated 22 patients with GK SRS and reported 95% tumor control rate and 67% rate of symptomatic improvement.

Pan et al evaluated outcome of GK SRS for 56 trigeminal schwannomas patients.²⁶ At a mean follow-up of 68 months, local control rate was 93% with resolution of neurologic deficits in 14 patients and improvement in 25 patients. Sun

et al reported outcome of SRS for 58 trigeminal schwannomas at a mean follow-up duration of 42.5 months.²⁷ They reported a tumor control rate of 93% with near disappearance of tumor in 7%, regression in 58%, unchanged tumor in 28%, tumor expansion in 7%, and the overall tumor control rate was 93%. Improvement in presenting neurologic symptoms was observed in 48%, stabilization of symptoms in 40%, and persistent cranial nerve dysfunction in 12% patients. Of the 13 patients who presented with secondary trigeminal neuralgia, 10 reported significant improvement.

Sheehan et al reported results of GK SRS for 15 trigeminal schwannomas.³⁰ At a mean follow-up of 4 years, they reported clinical improvement in 72%, a stable lesion in 16%, worsening of the disease in 12%, imaging documented regression in 48%, unchanged lesions in 40%, and increase

Table 2 Outcome of stereotactic radiosurgery for trigeminal schwannomas

First author	Year	Patient number	Mean follow-up (mo)	Median tumor volume (mL)	Margin dose (Gy)	Tumor control	Neurologic Symptoms	
							Better	Worse
Huang et al ²⁴	1999	16	44	5.3	15.3	100%	31%	0%
Nettel et al ²⁵	2004	23	40	4.5	15	91%	52%	9%
Pan et al ²⁶	2005	56	68	8.7	13.3	93%	68%	7%
Sun et al ²⁷	2006	58	42.5	4.6	13.1	93%	48%	12%
Peker et al ²⁸	2007	15	61	4	16	100%	40%	7%
Hasegawa et al ²⁹	2007	37	54	10	14.2	86%	40%	14%
Sheehan et al ³⁰	2007	25	48.5	3.9	15	88%	72%	12%
Phi et al ³¹	2007	22	37	4.1	13.3	95%	67%	27%
Kano et al ³²	2009	33	72	4.2	15	87.90%	33%	9%
Sun et al ³³	2013	52	61	7.2	13.9	86.50%	67%	4%

in size in 12%. Hasegawa et al reported outcome of GK SRS in 37 patients with trigeminal schwannomas and documented 10-year tumor control rates of 84%.²⁹ Peker et al treated 15 patients with newly diagnosed or residual/recurrent trigeminal schwannoma with GK SRS and reported 100% tumor control with tumor regression in 87% and stable tumor in 13% at a mean follow-up of 61 months.²⁸ Only one patient had transient facial numbness and diplopia. Kano et al retrospectively reviewed 33 consecutive patients with trigeminal schwannoma treated with GK SRS.³² Sun et al treated 52 patients with trigeminal schwannomas with radiosurgery using rotating gamma unit.³³ They reported a tumor control rate of 86.5% with near-total resolution in 15.4%, regression in 61.5%, unchanged tumor in 9.6%, and enlargement in 13.5% patients.

In most radiosurgery series no difference in outcome has been noted for patients who underwent primary versus adjuvant SRS for residual or recurrent tumor. The natural history of schwannomas in NF2 is different and NF2 cases cannot be managed like sporadic cases. The diagnosis of NF2 has been associated with poor outcome. In a study by Yianni et al, the 5-year progression-free survival was 96.2% for sporadic and 50% for NF2-related trigeminal schwannoma (TS).³⁵

Radiation Tolerance of Surrounding Structures

Dose selection for lesion near brainstem is essentially a balance between the anticipated therapeutic benefit and risk of an adverse radiation effect. We examined brainstem tolerance in a cohort of patients who underwent radiosurgery for intra-axial benign brainstem lesions. Seven (18.4%) patients developed adverse radiation imaging effects (ARIE).³⁶ ARIE correlated only with the presence of new neurologic deficits and age younger than 40 years. Three (7.9%) patients developed minor residual deficits without any ARIE. This study suggested that exposure of the brainstem to more than 12 Gy at volumes as low as 0.1 cm³ can produce ARIE and new neurologic deficits. The tolerance of the brainstem to radiosurgery is related to patient age, lesion volume, and pathology.

New cranial nerve deficit related to adverse radiation effect from after trigeminal schwannoma radiosurgery is rare. The motor nerves of the cavernous sinus (the third, fourth, and sixth cranial nerves) appear to be relatively resistant to radiosurgery.

Patient Selection for Radiosurgery

Patients with small- to moderate-sized tumors with intact cranial nerve function are optimal candidates for radiosurgery. Radiosurgery is also a good option for patients with symptoms related to cranial nerve dysfunction because these often improve after radiosurgery. For larger tumors initial resection followed by delayed radiosurgery for residual tumor is an effective option. GKS can be a reasonable alternative to surgical resection in patients harboring trigeminal

schwannomas without a severe compression of the brainstem. Radiosurgery is not an option for large volume tumors (tumor volume ≥ 10 mL) causing symptomatic mass effect on brainstem, tumors with deviation of the fourth ventricle, and tumors with brainstem edema. The tumors touching and impinging upon brainstem can be treated (as long as 12 Gy dose to brainstem is limited).

Fractionated Radiation Therapy

There are limited data on use of fractionated stereotactic radiotherapy for trigeminal schwannomas. Wallner et al described eight patients with nonacoustic schwannoma treated with postoperative radiation therapy from 1945 through 1983.³⁷ Irradiation was delivered 5 days per week at 1.6 to 1.8 Gy/fraction to a minimum tumor dose of 45 to 54 Gy. Two of the three patients who received postoperative irradiation following STR recurred. One of the three patients who received postoperative irradiation following NTR recurred. Two patients were treated with postoperative irradiation following biopsy and one recurred. Postoperative fractionated radiation did not prevent tumor recurrence after incomplete resection in nearly 50% of patients. Zabel et al reported 13 patients with nonacoustic schwannoma who were treated using fractionated stereotactic radiotherapy. Seven of these had trigeminal schwannomas.³⁸ Tumor size remained stable in nine and decreased in four patients. Two patients had a slight increase in tumor size at 3-month follow-up. In one of these cases, tumor regressed after 6 months, and in the other it remained stable. Temporary fatigue was seen in two patients and local erythema and temporary irritation of taste in one. At last follow-up, preexisting neurologic deficits remained unchanged in eight patients and improved in four. One patient complained of a slight increase in preexisting trigeminal hypoesthesia. Nishioka et al studied 17 patients with nonacoustic schwannoma who were treated with stereotactic radiotherapy between July 1994 and December 2006.³⁹ Five of these patients had trigeminal schwannomas. Radiotherapy was used as an initial treatment without surgery in 10 patients (59%) and after initial subtotal resection in the remaining patients. The tumor volume ranged from 0.3 to 31.3 mL (mean 8.2 mL). The treatment dose was 40 to 54 Gy in 20 to 26 fractions. Tumor regressed in 3 patients, remained stable in 13, and increased in 1 after SRT. One patient underwent resection at 32 months after irradiation.

The experience with fractionated radiation therapy for trigeminal schwannoma patients is very limited. Fractionated stereotactic radiotherapy may be an option for patients with large tumor who are higher risk for microsurgical resection and not a candidate for SRS.

Discussion

Management of Trigeminal Schwannomas

Observation, surgical resection, and radiosurgery are the cornerstone of management for patients presenting with trigeminal schwannomas. As in other specialties of medicine,

the “one size fits all” approach is slowly giving way to a more individualized treatment plan based on patient and tumor characteristics. Similarly, the “management by a single surgeon” is giving way to the “management by a skull base surgical team” to provide maximum expertise in both transcranial and transnasal approaches as well as radiosurgical and microsurgical strategies. In a treatment paradigm where complications such as brainstem damage, temporal lobe contusion, and CSF leak can be classified as present or absent, but outcomes such as facial numbness, diplopia, nasal crusting, temporomandibular joint pain, or headaches are more difficult to quantify, determining the exact value of each treatment modality can be challenging and often influenced by a patient’s preoperative condition and postoperative expectations. Radiosurgery appears to have the lowest incidence of new postoperative deficits, but the tumor control rate slowly declines with longer observation periods.

Most patients who present with partial fifth nerve involvement show symptoms improvements after SRS. Pan et al reported that after GKS 52 (93%) of 56 patients were successfully treated, with resolution of neurologic deficits in 14 patients and improvement in 25.²⁶ In a study by Sun et al, improvement of presenting neurologic symptoms was observed in 48%, stabilization in 40%, and deterioration in 12%. Among 13 patients with secondary trigeminal neuralgia, 10 had significant improvement or disappearance after GKS.²⁷ Sheehan et al reported clinical improvement in 72%, a stable symptoms in 16%, and worsening in 12%.³⁰ Kano et al documented neurologic improvement in 33.3%, stable symptoms in 57.6%, and progression in 9.1%.³² In a study by Sun et al, the rate of improvement of presenting neurologic symptoms was 67.3%.³³

Radical surgical resection provides the best long-term control rate at the price of the highest incidence of neurologic deficits. Size obviously matters in terms of treatment strategies and often strongly influences the initial recommendation. The role of a multidisciplinary skull base team is to help the patient go beyond the oversimplified strategy or “radiate the small ones and resect the big ones” to design a treatment paradigm that takes into account all the additional factors detailed earlier. With the quality of the current imaging, a very detailed analysis of a tumor and its surroundings should help better estimate the expected outcome from each treatment modality. A small tumor located on the cisternal portion of the trigeminal nerve, a few millimeters in front of the brainstem, and close to petrous bone’s trigeminal impression would be a more favorable radiosurgical candidate than the same size tumor, located on the second division of the trigeminal nerve, directly in contact with the mesial temporal structures on the dominant hemisphere. The cisternal segment tumor would be a more challenging reach with either a transcranial or a transnasal approach with a minimal expected complication rate from radiosurgical treatment while radiation damage to the dominant mesial temporal structures would be more of a concern for a V2 tumor that could be safely removed with minimal complications via either a middle fossa or a transnasal approach.

When given the option, a patient is most likely to favor first a “surgery without cutting” such as radiosurgery, then a “surgery without opening my head” such as endonasal approaches, before agreeing to a “real brain surgery” such as an open approach. An expert skull base team should be able to provide precise counseling for each patient’s situation to either reassure the patient that their initial choice is indeed the most appropriate or to diffuse irrational fears of a different strategy if a more invasive approach is indicated. The combination of surgery and radiosurgery for larger lesions cannot be overemphasized as a way to maximize quality of life and tumor control rate even in the most challenging cases. Finally, observation, although currently not advocated or supported by published series, should carefully be considered for a tumor that, histopathologically, has a benign appearance and might have a natural history comparable to that of vestibular schwannomas.

Conclusion

Improvement in neurologic symptoms, preservation of cranial nerve function, and control of mass effect are the primary goals of management for trigeminal schwannomas. An individualized management strategy by a dedicated skull base team with expertise in open and endoscopic approaches as well as radiosurgery should give the patient the best chance of achieving this goal. Where appropriate, a complete surgical resection is the treatment of choice, but this may not be possible in all cases. Radiosurgery is an option as primary management for small- to moderate-sized tumors and can be used for postoperative residuals or recurrences. For larger trigeminal schwannomas, initial surgical resection followed by SRS for residual tumor is an effective option, is associated with fewer cranial neuropathies, and greatly improves the patient’s overall outcome. The choice of endoscopic approach should be reserved for patients who have an extradural tumor or those isolated to the Meckel cave. The endoscopic approach is a minimally invasive approach with maximum benefits for a successful resection in select cases.

Note

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