Acoustic Neuroma: Viewpoint—Surgery

24

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Introduction

Acoustic schwannomas are the most common neoplasms arising in the cerebello-pontine angle. Treatment paradigms for patients with acoustic schwannomas may be inherently complex in nature, because of the multimodality treatment options available and numerous clinical considerations, including overall safety, hearing preservation, and facial nerve function, among others. Over the past several decades, the decision-making process for treating patients with acoustic neuromas has taken on even more complexity, given trends toward improved radiosurgical treatment modalities and earlier detection with advanced neuro-imaging. In experienced hands, surgical results provide rapid and reasonable success rates to those seeking an excisional cure. On the other hand, as longer-term follow-up has become available, radiosurgical tumor control data, both fractionated and unfractionated, have shown more impressive safety and tumor control rates. In selected cases, simple observation may also be a preferred strategy. Clearly, there is no best way to treat all patients with acoustic schwannomas, and various treatment strategies should be optimally used in a complimentary fashion to achieve the best overall outcomes.

In the ideal situation, a simple comparison between efficacy and safety data from several different therapeutic options should be sufficient to make an informed decision. However, aside from the various treatment options that exist, additional factors beyond effectiveness often come into play, including patient preference, surgeon bias, cost, patient age, and lifestyle issues. In our practice, the roles of providing recommendations for neurosurgical treatment and/or stereotactic radiosurgical treatment of acoustic neuromas depend primarily on four major factors: (1) patient age (2) tumor size (3) hearing levels, and (4) recurrence.

Conservative Management

As the availability of neuro-imaging studies increases, many patients will have new tumors diagnosed in an incidental fashion, without any subjective clinical symptoms. If tumors are small, a logical strategy may be to simply follow with surveillance imaging.

There are several studies, both short and long term, underscoring the fact that some tumors will change very little over time [1–5]. Depending on the study, up to 85 % of follow-up cases may show little or no growth of acoustic neuromas. In addition, several authors have demonstrated decreased tumor volume over time. Annual diameter growth rates have ranged between 0.15 and 4 mm [6–9]. Some practitioners, however, have witnessed alarming growth in rare cases, sometimes resulting in hydrocephalus and/or brainstem compression, highlighting the unpredictability of a strategy that relies simply on observation [5]. Cystic schwannomas, in particular, have been reported to have faster growth rates and present with more concerning signs related to brainstem compression [10].

Conservative management for young patients may therefore be fraught with hazard. More often than not, their tumors will grow, thus exposing them to greater risk associated with treatment, or even eliminating the potential for treatment alternatives. We have a large number of patients over the age of 70 who are simply examined on a yearly basis and undergo imaging. These patients infrequently require intervention, and radiosurgery is often employed if they do [11, 12]. Tumors that are too large to be treated with radiosurgery in patients within this age group are almost always offered operative therapy, unless there are substantial comorbidities that preclude general anesthesia.

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Surgical Management

The gold standard for treatment of any benign tumor is curative total removal. The concept of complete removal without the need for lifelong surveillance is appealing to many patients, making patient preference a major factor in the selection of primary therapy. Thus, when deciding between radiosurgical versus surgical alternatives, the realistic expectation of a safe and total surgical obliteration is a powerful incentive. Long-term follow-up data are available for selected surgical series documenting the potential for long-term freedom from recurrence in experienced surgeons' hands [9, 13–15]. In 379 surgical cases performed by the senior author, no recurrences resulted when the nerve of origin of the tumor was identified and sacrificed. Surgical treatment of acoustic schwannomas is often the preferred intervention in patients with larger tumors (>3 cm diameter), younger age, prior radiosurgical treatment, those with a preference towards surgical excision.

Prior to the advent of radiosurgery, it was the authors' philosophy to offer microscopic total removal to all patients with acoustic tumors. This strategy mandated, in many cases, protracted dissection of a thinned out facial nerve at the porus acousticus. Review of the senior author's facial nerve results shows 19 % of cases with long-term House Brackmann scores of 3 or worse. Given the high rate of radiosurgical control of small lesions, subtotal removal in certain narrow circumstances may be a preferable alternative.

Over the past two decades, however, a new strategy has been adopted in a majority of patients, in which the shift of the operative strategy focuses on aggressive tumor debulking with a major emphasis on maintaining facial nerve function. Depending on the surgical approach utilized, it is not uncommon to deliberately leave a residual amount of microscopic tumor along selected functional anatomical regions, such as the facial nerve, brainstem, or within the internal auditory canal (during retrosigmoid approaches). This strategy is most often employed in very large or vascular tumors. Frequently, no evidence of residual tumor is identifiable on postoperative MR imaging. If a visualized region of residual tumor is identifiable on postoperative MRI, this can be subsequently treated with radiosurgery, especially in cases where documented tumor growth of the residual component is observed [16, 17].

Long-term tumor control or cure is desirable to many patients, but at what price? Among larger series, surgical complication rates have become acceptably low. The most common complication is CSF leaks, occurring in 2–25 % of cases [9, 13–15, 18–20]. Major morbidity, including lower cranial nerve palsy, ataxia, or long-tract dysfunction, is also low, occurring 0–2 % of cases [9, 13–15, 18–20]. Mortality from surgery is rare, occurring in 0.4–2 % of patients. The two most important factors relating to safety and efficacy associated with acoustic neuroma surgery are tumor size and the surgeon's experience.

Facial Nerve Preservation Outcomes

Of all potential negative factors, a majority of patients are concerned about facial nerve function following surgery. In fact, this is clearly how the most accomplished practitioners of the surgical art keep score. It is important for the surgeon advising a patient to be familiar with statistical outcomes from recent surgical series and deem whether he or she can offer similar results. Most reports define an "excellent" or "good" outcome as a House-Brackmann facial nerve score of 1 or 2 [21].

A review of the senior author's results in 379 acoustic tumors of all sizes revealed a 79 % H-B 1 or two facial nerve function rate. Depending on the size of the tumor, excellent facial nerve functional results have been reported in up to 90 % of cases [9, 13–15, 22–30].

The Acoustic Neuroma Registry, started in the late 1980s, catalogues the national results of various surgeons through questionnaires and surveys. In a summary of 1,579 cases recorded in this registry between 1989 and 1994, Wiegand et al. found that in 92 % of cases surgeons felt the removal to be total with 94 % preservation of the facial nerve continuity [20]. Of these, 69 % had excellent facial nerve function after follow-up at 1 year.

In order to attempt a more direct comparison between surgery and radiosurgery, we sifted through the various publications to identify those who stratified facial nerve results based on tumor size. Facial nerve outcomes after surgery for tumors less than 3 cm were tabulated as single fraction radiosurgery only addresses this subgroup of patients. As expected, these facial nerve outcomes are laudable, with favorable facial nerve outcomes in the 80–99 % range [13–15, 25, 30, 31].

Another way to capture surgical results that can be compared with radiosurgical data is to look at cases where the surgeon attempted to preserve hearing. The majority of these lesions would be in the size range for single fraction radiosurgery. Shelton et al. reported that 89 % of their patients experienced good facial function in a series of 106 cases, performed through the middle fossa approach [32]. Other groups have elevated their surgical techniques to where facial nerve preservation and cochlear nerve preservation are an expected outcome [33–38].

Hearing Preservation

The data on hearing is a bit difficult to interpret as patient outcomes are sometimes reported as simply at or near pre-op levels. However, per the Gardner–Robertson scale a class I or II is considered as serviceable or better hearing [39]. In this scale pure-tone average of 0–30 db and speech discrimination of 70–100 % corresponds to class 1 hearing. Class 2 hearing assumes 31–50 dB pure-tone average and 50–69 % speech discrimination. The AAO-HNS Guidelines on the evaluation of hearing preservation also developed a system of classification with class A or B generally indicating useful hearing [40]. Class A hearing includes <30 dB pure-tone thresholds and >70 % speech discrimination. Class B includes 30–50 dB pure-tone thresholds and >50 % speech discrimination. Most reports use one of these two methods for recording hearing outcomes.

The data with regard to hearing preservation among the large series is somewhat variable. Good hearing can range from 24 to 60 % depending on the series [9, 13–15, 18, 25, 26, 28, 30, 31, 36]. It is noteworthy that the Acoustic Neuroma Registry data described by Wiegand et al. reports a 22 % hearing preservation rate [20].

A cursory look at data from groups who report large series of middle fossa approaches would suggest that this strategy might be more effective in terms of hearing preservation. Most accomplished groups report good hearing rates on the range of 50 % [32–35, 38, 41–43]. However, these series are all biased towards the smallest of tumors.

Surgical Approaches

Each of the three traditional approaches to the CP angle has their own putative strategic advantages (Table 24.1). The translabyrinthine approach minimizes retraction, maximizes exposure of the internal auditory canal, and facilitates defining the facial nerve within the canal. The retrosigmoid approach has the advantage of being stereotypical for most neurosurgeons while enabling the preservation of the hearing apparatus (Fig. 24.1). The middle fossa approach, while admittedly for smaller tumors, also facilitates exposure of the lateral most extent of the internal auditory canal. Proponents of each approach are able to post impressive numbers in terms of safety and cure rates. Many studies have looked at outcomes based on approach and no discernible advantage has been documented for one approach over another [15, 33]. However, for purely intracanalicular tumors several reports have shown that the middle fossa approach is safer for hearing preservation than the retrosigmoid approach [25, 35, 43]. A cursory review of the literature would suggest no way to resolve controversies that may emerge as to the best approach for a given situation. Hearing preservation, facial nerve function, and incidence of total removal seem to be practitioner or team related as opposed to approach related. This suggests that technical expertise and strategic operative decision-making can negate most disadvantages related to the various approach strategies.

Table 24.1 Approach selection.

	Retrosigmoid	Middle fossa	Translabyrinthine
Size			
<1 cm			
Lateral impact	+	+++	++
Medial	+++	+++	++
<2.5 cm	+++	0	+++
>2.5 cm	++	0	+
Only-hearing ear	+++	0	+
Hearing			
>50/50	+++	+++	+
<50/50	+	+	+++
Recurrent	+	0	+++

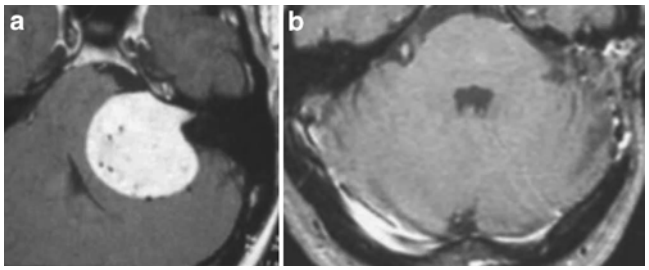

Fig. 24.1 (**a**) A 5 cm acoustic neuroma prior to resection. The patient presented with tinnitus, hearing loss, and signs of brainstem compression. (**b**) Postoperative image after a retrosigmoid craniotomy. All preoperative symptoms resolved

A retrospective review of our institution's last 50 cases of retrosigmoid resections was compared to the last 50 translabyrinthine resections. Tumors were included that were 3 cm or less and results were categorized in terms of facial nerve outcome, incidence of total resection, and major complications. At discharge, 37 of the retrosigmoid patients had an H-B score of 1 as compared to 28 of the translabyrinthine patients. Combining H-B 1 and 2 scores showed 82 % with the retrosigmoid cases and 70 % of the translabyrinthine. This did not reach statistical significance. There was one major neurological complication in the retrosigmoid group and none in the other group. Three cases of subtotal removal resided in each group. Long-term follow-up of these patients showed even less of a difference in facial nerve function. This suggests that surgical technique may be more important than approach strategies in terms of outcome for acoustic tumors.

Ideally, each practitioner should have a working knowledge of all approaches. This can instill the wary patient with some confidence that decision-making is based on assess-

ment of the patient's best interests. For those who are experiencing unacceptable results, altering approach strategy may show modest benefits, but doing such will not make up for deficiencies in surgical technique as it relates to the brainstem vascularity and cranial nerves.

Choice of Treatment Method (Radiosurgery Versus Excision)

At our institution, the three traditional approaches for surgical excision of acoustic tumors are practiced. Further, additional technologies for radiosurgery (such as Gamma Knife, Linac, Cyberknife, etc.) are offered. The surgical decision-making is less predicated on available technologies or reliance on a practiced single surgical approach, than it is on patient age, tumor size, and need for hearing preservation (Table 24.2).

Age and Tumor Size

For younger patients, emphasis is placed on surgical removal. A surveillance strategy in this group is likely to be futile, since inevitably the lesion will cause further symptoms and require treatment. Radiosurgery has a long follow-up period and the window of vulnerability for recurrence is potentially wide. Reliable data on lifelong tumor control for patients in their 30s or 40s is lacking. For those in their 50s and 60s, single fraction radiosurgery is an attractive alternative. Efficacy and safety statistics are available for this group and are highly acceptable (Fig. 24.2). Patients in older age groups rarely need any therapy unless their tumor is large enough to be threatening [11, 12]. Furthermore, radiosurgery is not without its complications and failures, which develop in approximately 3–34 % of cases in even the most experienced centers [44, 45]. Serviceable hearing is lost in 30–43 % of patients following radiosurgery [46, 47]. Finally, in the event that surgical excision is required following radiosurgery, it has been reported that tumor excision is much more challenging, and the resiliency of the facial nerve is compromised, compared to nonirradiated patients [48].

Table 24-2. Treatment modality.

	Radiosurgery	Surgery	Observe
Size			
>2.5 cm	+	+++	0
1.5 to 2.5 cm	++	++	+
<1.5 cm	+++	++	++
Age			
<40	+	+++	0
40 to 60	++	++	+
>60	+++	+	+++
Hearing			
>50/50	+++	++	++
<50/50	++	++	+
Recurrent	+++	+	+

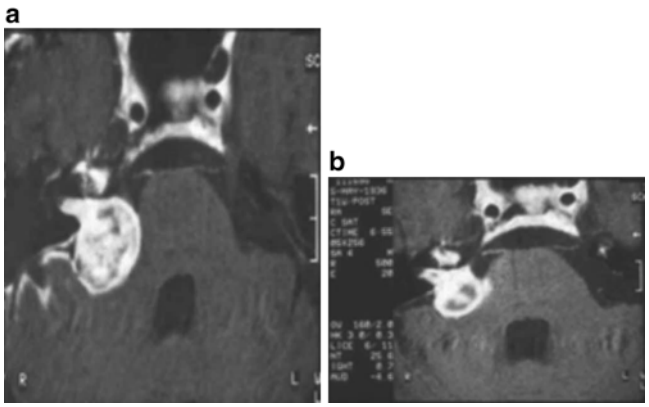

Fig. 24.2 (**a**) A 2.5 cm acoustic neuroma in a patient who presented with slight hearing loss. The fourth ventricle is distorted. This patient elected to have radiosurgery. (**b**) Post gamma knife radiosurgical treatment. Central necrosis is present as well as decreased pressure on posterior fossa structures

For large lesions greater than 3 cm, single fraction radiosurgery has no role [49]. For these lesions, gross surgical removal or in certain situations subtotal removal, with follow-up radiosurgery or observation, is advisable. Subtotal removal for a patient with a single tumor and good hearing in the other ear should be an unusual event. A small residual may be left behind in an effort to avoid a major complication such as facial nerve sacrifice or brainstem injury. For younger patients with large tumors, surgical removal is the preferred strategy. The length of vulnerability for recurrence is too great for younger patients to rely on subtotal removal. Some further therapeutic endeavor will ultimately be necessary, multiplying the potential for complications.

Large lesions in older patients can present some strategic problems. This would seem like an ideal situation for hypofractionated radiosurgery. However, with lesions greater than 4 cm or somewhat smaller lesions with associated arachnoid cysts usurping much of the available reserve in the posterior fossa, radiosurgery with its attendant edema formation may produce unacceptable risks 6–12 months posttreatment. Data is sorely lacking for this modality in larger tumors. Until better long-term studies are available, older patients in good health with large lesions should be offered the option of surgical removal. The decision for total versus subtotal removal is made at the time of surgery and predicated on the likelihood of complications. Radiosurgery as an adjunct can be offered for any threatening residual. For large tumors in older patients who are poor surgical risks, hypofractionated radiosurgery is a logical option.

Hearing Preservation

Hearing preservation in the context of surgical removal can be expected in experienced hands to be successful 50 % of the time with intracanalicular tumors and 30 % with larger lesions that are generally under 3 cm. This presupposes good functional hearing to begin with. Attempts to save hearing in a marginal or poorly hearing ear will be unrewarding. That ear will be a constant source of distraction to the patient as it picks up unstructured background noise and reduces the overall functionality of the hearing. Thus, compromises in surgical strategy to preserve the function of a poorly hearing ear should be vigorously resisted.

Single fraction radiosurgery is fast growing in popularity and may become the treatment of choice in the absence of an experienced surgeon with a proven track record for the safe and effective removal. For those lesions less than 3 cm, one can expect at least 50 % hearing preservation or better assuming accepted proven radiosurgical techniques are utilized. The major drawback in prescribing it for all small acoustic tumors is the lack of long-term efficacy data. For many patients, the need for continued surveillance and the thought of the continued presence of the lesion are negative satisfiers.

If an acoustic neuroma recurs and is deemed in need of treatment, the first option should be radiosurgery. This assumes the tumor regrowth has been detected before it has grown too large for radiosurgery. In these cases a translabyrinthine surgical approach will offer the largest corridor while minimize the need for retraction or dissection of previously scarred brain. A repeat retrosigmoid craniotomy may prove difficult.

Choice of Surgical Approach

Our preference is to use the translabyrinthine approach for all tumors where hearing preservation is unlikely (Table 24.1). Thus, it is used in all large tumors and those with poor hearing. Certainly those greater than 3 cm would be treated this way and most with speech discrimination scores of less than 50 %. The only time we would favor a retro sigmoid approach in a large tumor would be the case of a large lesion in an only hearing ear where a subtotal removal is contemplated.

In tumors that protrude from the porus acousticus, the retrosigmoid approach is preferred for hearing preservation (Fig. 24.3). In our most recent 80 cases using this approach, functional hearing resulted in 30 %. We limit the use of the middle fossa strategy for those small intracanalicular lesions that are impacted in the lateral end of the internal auditory canal.

Other factors come into play as patients try to make informed decisions. Socioeconomic and educational status may complicate decision making in patients who cannot understand a complex set of options. Patient and family biases for or against surgery or radiation may direct the patient's thinking contrary to the physician's best judgment. Access to the internet, influence from patients who have had one form of therapy or another, and loyalty to a particular institution may be relevant factors in decision-making.

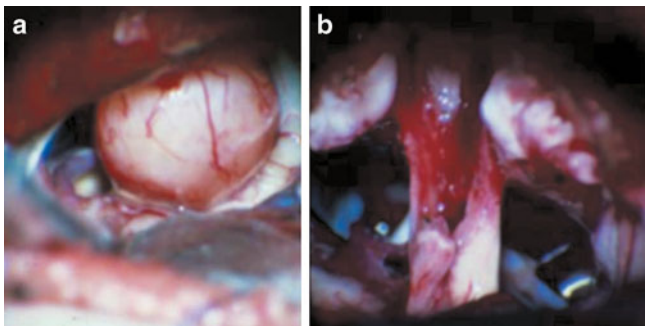

Fig. 24.3 (**a**) Intraoperative view of acoustic neuroma during a left retrosigmoid craniotomy. (**b**) After tumor removal, the nerves of the porus acousticus are seen. Note to transected superior vestibular nerve from where the tumor originated

One can guide the decision making by trying to simplify principles. Explaining away misconceptions is a place to start. Identifying patient and family biases and dealing with them in a forthright way will also help. If it is perceived that this discussion is simply a device to steer the decision-making toward the surgeon or radiosurgeon's bias, confusion and mistrust can develop. If a patient harbors a tumor that may be amenable to either surgical removal or radiosurgery, a simple construct can be presented to the family and patient to facilitate their decision-making. Does the patient insist that the tumor be gone? Benefits include diagnostic certainty and the lack of need for long-term surveillance.

Ultimately patients will decide on what treatment option they prefer with some guidance as to the risk/benefit ratio from the physician. The surgical removal of acoustic neuroma has been refined over time to achieve impressive results. However, obvious risk of major morbidity remains. Radiosurgical methods are progressively less invasive and less likely to cause major morbidity. However, large lesions are clearly not amenable to this therapy and lifelong observation is a requirement, even for small lesions, and especially in younger patients. No perfect algorithm exists and each patient scenario has unique challenges. The first question the physician and patient must answer is how important is the removal of the tumor to one's overall comfort. If knowing the tumor is still present makes one unable to participate in life activities then surgery seems logical. Otherwise the radiosurgical alternatives must be explored. With sound advice and research, the patient will ultimately decide his own treatment algorithm.

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