Technical nuances of resection of giant (> 5 cm) vestibular schwannomas: pearls for success

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Removal of vestibular schwannomas (VSs, or acoustic neuromas) remains one of the most challenging operations in neurosurgery. Giant or huge tumors (> 5 cm) heighten these challenges, and technical nuances play a special role in maximizing tumor resection while minimizing complications. In this article, the senior author describes his technical experience with microsurgical excision of giant VSs. The accompanying video further illustrates these details.

Key words: • acoustic neuroma • vestibular schwannoma • technical nuance • complication • giant tumor

Acoustic neuromas (or VSs) are one of the more technically challenging neurosurgical lesions to remove as exemplified by the well-demonstrated career-long learning curve in VS surgery.6,23 There is a long-recognized difference in outcome between low- and high-volume surgical centers.31 Giant VSs, greater than 4–5 cm in greatest extracanalicular diameter,12 carry their own unique risks for resection. Tumor size has been repeatedly shown to correlate with patient outcome, including postoperative cranial nerve and brainstem function.11 Higher complication rates2,28 have been reported with large (> 2 cm) and giant tumors. However, in the hands of experienced surgeons, giant VSs can be removed safely with no deaths, low morbidity, and cranial nerve preservation.24 In this paper we describe the nuances of the preoperative, intraoperative, and postoperative management of giant VSs framed in the context of the case presented in the accompanying video (Video 1).

Video 1. Clip showing removal of a large VS (> 5 cm). Technical nuances play an important role in maximizing tumor resection while minimizing complications. This video of an operation performed by Aaron A. Cohen-Gadol, M.D., M.Sc., illustrates these details. Click here to view with Media Player. Click here to view with Quicktime.

Abbreviations used in this paper: IAC = internal auditory canal; VS = vestibular schwannoma.

Preoperative Evaluation

All patients should be evaluated clinically with a complete history and neurological examination focusing on cranial nerve and brainstem/cerebellar function, as well as long tract signs: most tumors of this size (> 5 cm) are Hanover Class T4A or T4B with significant brainstem compression. The most common presenting symptom is significantly decreased or absent ipsilateral hearing. In patients with residual useful hearing, formal audiometry should be performed because preservation of hearing is reported to be possible occasionally and may influence the surgical approach. Pure-tone audiograms and speech discrimination testing is the standard, with results classified according to a number of methods, commonly the Gardner-Robertson scale.9 Auditory brainstem response screening plays a little role, given the overt clinical symptomatology at presentation. Other common cranial neuropathies in recent series of giant VSs include tinnitus (34%–100%); extraocular muscle impairment (4%, usually abducens nerve dysfunction); trigeminal sensory dysfunction (4%–17%); facial palsy (1.6%–14%), which should be objectively evaluated using the House-Brackmann scale; and dysphagia/lower cranial nerve dysfunction (4%–5%), which if significant, merits formal swallow and vocal cord mobility studies. Signs of cerebellar or brainstem compression may also be present, including ataxia (also related to vestibular dysfunction, 28%–64%), dysmetria (7%), hyperreflexia (14%),
as well as signs and symptoms of intracranial mass effect or hydrocephalus (7%–33%).\(^{2,3,25,30}\)

An important consideration is the management of hydrocephalus. Preoperative untreated hydrocephalus has previously been connected to poorer outcomes and higher complication rates.\(^{18,34}\) The majority of patients do not require further treatment for their hydrocephalus besides tumor resection; however, a minority do not achieve such a goal and may require postoperative ventriculoperitoneal shunting. As expected, increased tumor size is a risk factor for development of postoperative hydrocephalus.\(^{37}\) If symptomatic obstructive hydrocephalus is present, we prefer to place an external ventricular drain at the time of craniotomy with the goal of weaning the drain postoperatively, if possible, and shunt placement if necessary. In the particular case described in the accompanying video, due to the patient’s gravid state and associated risks, an initial shunting procedure was performed to temporize the patient’s symptoms until postpartum to allow for delayed tumor resection.

**Imaging Studies**

Imaging studies should include CT and MRI. Fine bone detail is important to evaluate the bone anatomy that will be removed during the creation of the surgical corridor. Recognition of the extent of pneumatization of the temporal bones is important to prevent postoperative CSF leak. The position of the venous anatomy, specifically a high-riding jugular bulb or posterior sigmoid sinus, may affect the choice and extent of bone removal.\(^{13}\) Tumor anatomy and its relation to neurovascular structures is best evaluated using MRI, which allows differentiation from other cerebellopontine angle lesions, such as meningioma or epidermoid.\(^{32}\) Outgrowth from the posterior fossa is critical to note, and extension of giant tumors around the tentorium may indicate the need to resect a portion of the tentorium for adequate tumor exposure and microdissection. Beyond the local anatomical relationships of the tumor, its intrinsic characteristics such as a cystic component can be identified, which has been correlated with poorer outcomes.\(^{8,26}\) The relationship of the tumor to the basilar and posterior inferior cerebellar arteries may be appreciated; such vessels should be carefully protected if ultrasonic aspirator devices are used for tumor decompression. Despite extension of these giant tumors through the jugular foramen, the tumor can typically be microsurgically dissected off of the lower cranial nerves with no significant risk, as exemplified by the low rate of postoperative deficit (2%–6%). Presence of edema in the brainstem indicates a high risk of brainstem pial violation during microdissection of the tumor capsule.\(^{19}\) If significant brainstem edema is evident, staging the surgery may be strongly considered, as the interval between 2 stages would allow the tumor to deliver itself into the resection cavity created during the first operative session.\(^{39}\)

Representative preoperative and postoperative MR images from the senior author’s patient are shown in Fig. 1, demonstrating the presence of minimal brainstem edema, extent of brainstem compression, and the presence of tumor within the jugular foramen.

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**Operative Preparation**

Staging the operative session should be strongly considered in giant tumors to provide the patient with the best outcome. Fatigue of the surgeon and the operative team during the later (and more critical) parts of the operation is an important factor.\(^{29}\) In a staged procedure, one accepts the risks of a second surgery for the benefits of shorter procedures that are less taxing for the patient and surgeon. Indeed, early series of staged surgery for these giant tumors demonstrated no deaths and acceptable morbidity (65% functional facial nerve preservation).\(^{29}\) Modern staged series have shown that comparable outcomes with minimal additional risk can be achieved via staged resection for large tumors, and have even suggested superior facial nerve outcomes.\(^ {4,17,19}\) Recent microsurgical and neuroanesthetic techniques weigh in favor of a single-stage surgery: modern series have achieved excellent results (<1% death, 70%–75% functional facial nerve preservation, 97%–100% excision).\(^{3,22}\) In a young, otherwise healthy patient such as the one described in the video, a single-stage surgery is a reasonable consideration.

Routine use of intraoperative facial nerve electromyography as well as brainstem auditory evoked response potentials and somatosensory evoked potentials to identify and monitor facial nerve and cochlear nerve/brainstem integrity, respectively, are important. We find these to be an invaluable guide to safe resection and for revising a surgeon’s maneuvers intraoperatively to prevent neural injury. It should be noted, however, that the significant attenuation and atrophy of the facial nerve in giant tumors, as apparent by preoperative facial weakness, often complicates mapping the exact location of the nerve splayed over the tumor capsule.

Various skull base approaches and their combinations are described for removal of giant VSs, including retrosigmoid suboccipital and translabyrinthine routes. Numerous experts have advocated for each route, generally based on their personal expertise, achieving excellent outcomes.\(^{3,14,25,30}\) Those experienced in both denote benefits to each approach and tailor their own approaches to each patient’s unique anatomy,\(^{13}\) while admitting that the best approach is often the one most familiar to the surgeon. Raslan et al.\(^ {19}\) recently described a 2-stage operative session for these tumors and employed both retrosigmoid and translabyrinthine approaches in each session with great results. The decision to stage tumor removal was made based on evidence of cerebellar or brainstem edema, significant tumor adherence to the brainstem and facial nerve, a poorly stimulating facial nerve (partial nerve injury), and attenuated facial nerve.

In the modern era, when the goal of zero deaths and minimal morbidity is achievable, hearing conservation may be considered; in large and giant tumors, this is the exception rather than the rule,\(^ {35}\) and only in the few cases in which serviceable hearing remains preoperatively. Venous anatomy should be considered; a high and/or anteriorly located sigmoid sinus significantly favors selection of a retrosigmoid approach.\(^ {15}\) In our experience, the translabyrinthine approach affords the opportunity for less cerebellar retraction and may be a consideration for
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younger patients with a “full” cerebellum. This route may be used in combination with the retrosigmoid route as it may not provide enough exposure of the brainstem for giant tumors. The retrosigmoid approach provides a more panoramic view of the compressed brainstem, however, and has generally been our preference. This approach allows for a single-stage resection of the tumor.

Patient Positioning

The patient may be positioned in sitting/semisitting or horizontal (supine, oblique, park bench) fashion. The sitting position is classically associated with a risk of venous air embolism; precordial Doppler and/or end-tidal CO₂ monitoring are used. Some operators report less blood loss, less operative time, and lower cranial nerve dysfunction with the sitting position. The sitting/semisitting position will facilitate a clear surgical field by using irrigation alone. Bimanual microdissection is a great advantage without the use of bipolar electrocautery, which could place the facial nerve at risk. The sitting position will place the arms of the surgeons at risk for fatigue. Nonetheless, surgeon’s preference and familiarity with a particular position is most important. We avoid the supine position due to the risk of neck stiffness associated with such long operative sessions. Our preference has been the park-bench position.

Intraoperative Nuances

For our retrosigmoid approach, a curvilinear incision (Fig. 2A) prevents the scalp flap from interfering with the working zone of the surgeon and decreases the operator’s working distance to the tumor (Fig. 2B and C). A large retromastoid craniotomy/craniectomy extending along the edges of the transverse and sigmoid sinuses is desirable. Smaller craniotomy/craniectomy may not provide adequate decompression if intraoperative cerebellar swelling is encountered. Bone removal for giant tumors extends to the posterior fossa floor, but does not involve the opening of the foramen magnum. If significant obstructive hydrocephalus is present preoperatively, CSF drainage through a previously placed external ventricular drain is achieved before dural opening to avoid cerebellar herniation. Dural opening is completed using a curvilinear incision (Fig. 3A). Additional CSF is released by opening the arachnoid membranes caudal to the tumor (Fig. 3B). A small lateral portion of the cerebellum may be excised to allow for adequate tumor exposure without aggressive cerebellar retraction; this may be necessary only in younger patients with a “full” cerebellum, otherwise cisternal opening and drainage may be adequate. The arachnoid membranes over the posterior aspect of the tumor capsule are excised.

Fig. 1. Representative axial MR images from the senior author’s patient, demonstrating the extent of brainstem compression (A), presence of minimal brainstem edema (B), and tumor within the jugular foramen (C). Postoperative images (D and E) reveal gross-total removal of the mass. Images A, C, and D are T1-weighted with contrast enhancement, images B and E are T2-weighted.
Stimulation of the posterior/inferior capsule will exclude an aberrant posterior/inferior displacement of the facial nerve (Fig. 3C). Coagulation of the posterior capsule is followed by aggressive internal tumor debulking (Fig. 4). Maximal internal debulking will tremendously facilitate the later stages of the operation due to enhanced tumor capsule mobilization from the surrounding cerebrovascular structures, as well as decrease neural tissue stretch/retraction. Following devascularization of the tumor by coagulating the feeders from the dura over the porus acusticus, microdissection continues inferiorly as the tumor capsule is mobilized away from the lower cranial nerves and out of the jugular foramen (Fig. 5A). This stage of microdissection should be atraumatic as the lower cranial nerves do not significantly attach to the capsule. Stimulation of the inferior pole of the tumor should exclude the presence of the facial nerve. Subsequently, the tumor is internally debulked along its superior pole and rostral dissection continues, mobilizing the capsule away from the tentorium and fifth cranial nerve. The facial nerve is often adherent along the superior pole of the tumor and adjacent to the trigeminal nerve. This portion of the capsule should be carefully mapped with the stimulator. Movement of the temporalis muscle caused by trigeminal nerve stimulation should not be mistaken for localization of the facial nerve. Mapping of the often attenuated/atrophied facial nerve in giant tumors can be difficult, especially in the presence of preoperative facial weakness. Repetitive mapping at slightly higher stimulation parameters may be necessary to completely exclude the presence of the facial nerve in the region. The capsule is sharply dissected away from the distal trigeminal nerve (Fig. 5B); the trochlear nerve and superior cerebellar artery are preserved. The superior petrosal vein is protected.
if possible. Venous bleeding from the petrosal-tentorial junction should be carefully controlled.

The tumor capsule is subsequently rolled laterally away from the middle cerebellar peduncle and brainstem toward the porus acusticus. Meticulous hemostasis will allow the operator to appreciate the most important factor in safe resection of these challenging tumors: microdissection along the arachnoid membranes and respecting the brainstem pial membranes. The capsule is pulled on gently as dissection is performed using microforceps to detach the arachnoid membranes from the tumor while periodic irrigation by the assistant clears the field. Suction over the brainstem and cranial nerves is strictly avoided. The veins along the surface of the brainstem are often engorged and prone to avulsion, leading to blood loss and interference with adequate visualization of the dissection planes.

Gentle pressure using a cotton ball over the site of the hemorrhage followed by coagulation of the vein along its more proximal segment away from the brainstem is a possible strategy.

Internal debulking using an ultrasonic aspirator followed by tumor mobilization is a safe maneuver to avoid inadvertent injury by undue retraction of the surrounding structures. Further mobilization of the inferior pole of the tumor should protect the posterior inferior cerebellar artery and its branches. En passage vessels are microsurgically mobilized using sharp dissection as these may be crucial vessels, while small tumor-feeding vessels are carefully coagulated and cut. Blunt dissection of the perforators and their subsequent avulsion must be avoided. Cranial nerve VIII is often encountered in the region; its preservation in giant tumors is almost impossible and not advisable if preoperative hearing is nonfunctional.

In the presence of preoperative brainstem edema, violation of the pial membranes is likely in the giant tumors. If such an event occurs, a small piece of cottonoid may be used to mobilize (peel away) the brainstem from the tumor (Fig. 5C) without placing the former at risk for injury by the suction apparatus. Additional cottonoid patties are added and left behind until the end of the operation, when they are irrigated away. The pial membranes may be reidentified along the inferior pole of the tumor. As the tumor is mobilized away from the brainstem along its superior pole and midsection, mapping will localize the facial nerve along the capsule or at its exit zone along the brainstem. The most reliable maneuver to expose the nerve safely is to peel the tumor laterally and identify the nerve along its root exit zone at the brainstem. Removal of the tumor in the deep cerebellopontine cleft may require the most amount of cerebellar retraction; changing the angle of the microscope’s view and intermittent dynamic retraction using the suction tip may minimize the required persistent force. The length of the operation, any change in vital signs, or significant violation of the pial membranes or concerning facial nerve recordings may lead the operator to stage the operation.

Localizing the facial nerve along the superior half of the capsule, the surgeon can be aggressive in removal of the inferior pole, which is often not very adherent to the brainstem; the abducent nerve is often adherent to the capsule. Continuing to roll the superior pole laterally, the surgeon will expose the root entry zone of the trigeminal
nerve. This part of the nerve is often very adherent and draped over the tumor, and careful dissection technique will minimize the risk of postoperative trigeminal neuropathy and resultant corneal anesthesia. Attentive internal decompression of the residual tumor will facilitate capsule mobilization using sharp dissection techniques. Using meticulous frequent stimulation, the facial nerve is dissected and peeled away from the tumor; mobilizing the tumor away from the nerve may place the nerve at an increased risk of injury (Fig. 6A–C). Bleeding can be a nuisance, but blind bipolar coagulation should be avoided. At this stage in a cooperative procedure, the neurosurgeon may take a break while the neurotologist removes the tumor within the IAC (Fig. 6D–F).

The inferior wall of the IAC is drilled to the fundus after the dura is dissected from the petrous bone. Cutting and diamond burs may be used to remove the bone under generous irrigation to avoid heat injury to the nerves in the canal; a large piece of soaked soft cotton is used to cover the cerebrovascular structures in the subarachnoid space to protect them from the bone dust during drilling. Extension of bone removal will allow identification of the distal cranial nerve VII/VIII complex free of tumor (Fig. 6E). The mastoid air cells will often be entered and should be carefully waxed at the end of this stage. The dura within the IAC is cut and the tumor is debulked and rolled medially. The vestibular nerve is meticulously identified and the facial nerve is mapped before the vestibular nerve is cut, to allow the tumor to be mobilized out of the canal (Fig. 6F). The facial nerve becomes very attenuated and adherent to the tumor along the junction of its subarachnoid and intracanalicular segments. The operator has to use a careful combination of gentle blunt and sharp microdissection techniques during tumor mobilization at the level of the porus to avoid facial nerve injury.

Removal of the tumor within the IAC will convey additional information regarding the route of the facial nerve over the capsule from its already identified root exit zone to the area of the porus acusticus. If the facial nerve is significantly attenuated, its anatomical preservation may not be possible in giant tumors. Although gross-total tumor resection is attempted, if the tumor is very adherent to the nerve at the level of the porus, a small piece of the tumor may be left behind to optimize facial function. This small piece of the tumor left over the nerve can be managed postoperatively through surveillance imaging and treated with radiosurgery (if enlarging) with good rates of tumor control. The surgeon may use the blunt-tipped stimulator as an instrument to peel away the nerve from the capsule. Significant tension on the nerve is avoided and sharp dissection is used (Fig. 7). Any injury to the nerve may require an increase in stimulation parameters to map the nerve. The basilar artery is evident at the end of the resection. Basilar artery perforators should be preserved. Meticulous hemostasis is followed by a watertight dural closure and generous application of bone wax to the mastoid air cells. If cerebellar swelling is evident, the bone flap should not be replaced and a generous suboccipital decompressive craniectomy is performed. Figure 8 illustrates the typical patterns of facial nerve displacement by large and giant VSs.

**Postoperative Considerations**

Patients are observed in the intensive care unit for
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any signs of neurological deterioration, sudden hypertension, or breathing and swallowing difficulty. Meticulous postoperative care is mandatory to detect and prevent complications. Appropriate eye care is provided: the combination of trigeminal neuropathy and facial nerve paralysis places the eye at risk and appropriate precautions are taken in the presence of one or both. If dysphagia or respiratory insufficiency is encountered, early percutaneous gastrostomy and tracheostomy tubes are placed to avoid any complications such as aspiration pneumonia and hypoxemia; the possibility of these is important to discuss with the patient preoperatively.

If a staged operation is planned, the interval between operations may be 2–4 weeks based on the patient’s recovery process from the first operation. Others have advocated for a longer interval to allow further cranial nerve and brainstem recovery. If poor facial function is present from the first operation, the second stage is delayed until the nerve achieves a good functional recovery.

Complication Management

The most feared complication with this operation is intraparenchymal hemorrhage and cerebellar edema, which contributes the main source of perioperative morbidity. Avoidance of significant retraction on the nervous structures intraoperatively, meticulous microdissection along the arachnoid membranes, and prompt management of postoperative hypertension will minimize these unfortunate events. Watertight dural closure is important to avoid pseudomeningocele formation. If rhinorrhea is encountered, we perform early mastoidectomy and obliterate the air cells with a fat graft.

The main neurological morbidity of VS surgery is facial nerve palsy, which is both functionally and psychologically damaging to the patient. This is especially true with giant tumors, as tumor size is the main factor predicting postoperative facial weakness. Patients with an incomplete eye closure may undergo gold weight placement within the eyelid and/or tarsorrhaphy. Facial reanimation procedures may be considered if facial nerve function does not return within 1 year postoperatively. This maneuver may be considered earlier if the nerve was anatomically noncontinuous at the time of surgery.

The goal of the surgery remains gross-total resection of the tumor and preservation of function. Meticulous microsurgical techniques remain the important factor in operative success.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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