Giant vestibular schwannomas: Surgical nuances influencing outcome in 179 patients

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ABSTRACT
Introduction: Giant vestibular schwannomas (VSs; ≥4 cm in size) are considered a separate entity owing to their surgical difficulty and increased morbidity. We studied the clinical presentation and surgical outcomes in a large series of giant VS patients. We also present the surgical nuances, which we believe can improve surgical outcomes.

Materials and Methods: The clinical profiles, radiology, surgical results, and complications of 179 consecutive patients with a unilateral giant VS were reviewed. The study population was classified into two groups: Group A (4–4.9 cm, 124 [69.3%] patients) and Group B (≥5 cm, 55 [30.7%] patients).

Results: The mean tumor size in Group A was 4.3 ± 0.2 cm (range, 4–4.8 cm), and in Group B, it was 5.3 ± 0.4 (range, 5–6.7 cm). Patients in Group B were younger, with a mean age at presentation of 34.8 ± 12.3 years versus 41.8 ± 13.1 years in Group A (P < 0.05). There was no difference in the clinical presentation except for papilledema (81.8% vs. 66.9%) and VI cranial nerve (CN) dysfunction (9.1% vs. 2.4%; P < 0.05), which was higher in Group B. There was no difference in the rate of total excision (86.2% vs. 85.4%), anatomical and physiological facial nerve preservation rates between the two groups (approximately 2/3 and 1/3, respectively), and the facial function at discharge. The incidence of postoperative morbidity was not statistically different between the two groups, except for the occurrence of postoperative cerebrospinal fluid (CSF) rhinorrhea, which was greater in Group B (10.9% vs. 2.4%). There were two mortalities in each group (overall, 4/179; 2.2%; P = 0.58).

Conclusions: Patients with ≥5 cm VSs were younger, with a higher incidence of papilledema and lateral rectus paresis. However, when compared with tumors ≥4 cm in size, there was no difference in the extent of excision, facial nerve preservation, and postoperative complications (except CSF rhinorrhea) or mortality. Thus, further subclassification of giant VSs does not seem to be necessary.

Key words: Brain tumor; giant, size; surgery; vestibular schwannoma

Introduction
Surgery for giant vestibular schwannomas (VSs; ≥4 cm in size) is a formidable challenge. Following advances in and due to widespread availability of brain imaging, the incidence of these tumors in the Western countries has reduced to approximately 2% of all VSs;[1] the incidence in Europe is approximately 12.5%; however, it remains as high as 50% in the Indian subcontinent.[2,3]

The clinical condition of patients with giant VSs differs significantly from patients with smaller tumors, and the surgical morbidity and mortality rates are higher in those with larger tumors.[4] While we know from previous studies that the outcomes of giant tumors are worse than that of...
tumors <4 cm in size,[5] it is unclear whether there would be a substantial difference in the presentation and surgical outcomes of giant VSs of different sizes. We conducted this study to determine the benefit, if any, in subclassifying giant VSs. We also wanted to document surgical nuances that might influence outcomes.

Materials and Methods

The demography, clinical profile, radiological features, and surgical and postoperative outcomes of patients who presented with unilateral VSs to our department between 2002 and 2008 were retrospectively analyzed. All information was retrieved from the medical records of these patients. Of the 318 patients operated for VSs, 179 (56.2%) who had a tumor size of ≥4 cm were included in the study. They were classified into two groups: Group A (4–4.9 cm, 124 [69.3%] patients) and Group B (≥5 cm, 55 [30.7%] patients). All patients underwent preoperative magnetic resonance imaging (MRI), and the tumor size was calculated using the single largest diameter in the subarachnoid space [Figure 1]. Tumors classified as predominantly cystic had a cystic component larger than half the tumor size. The facial nerve function was graded according to the House-Brackmann (H and B) grading system,[6]

Preoperative shunt

Preoperative shunts were not performed routinely even in patients with clinical features of raised intracranial pressure with imaging showing hydrocephalus. Those who received shunts had already had these done prior to presenting to us; those who had significant impairment of vision with impending visual loss; and, those who had the possibility of waiting for a definitive surgery due to logistical reasons.

Surgical technique

Although multiple surgeons preferring different operating positions performed the surgeries, the microsurgical techniques were largely similar. Patients were operated via the retrosigmoid suboccipital approach either in the supine–oblique, lateral, or semi-sitting position. Intraoperative facial nerve monitoring was routinely used. As no patient had a preoperative serviceable hearing on the side of the lesion, no attempt was made to preserve hearing at surgery. Our philosophy was to excise tumors completely, as patients came from distant parts of the country and were unlikely to return for follow-up, and most patients often wanted a “permanent cure.” However, when patients expressly requested that facial nerve function be preserved at the cost of a subtotal or near-total excision, we adhered to their request. In some patients even without a preoperative expressed desire for facial nerve preservation, the surgeon chose to leave some bits of the tumor on the facial nerve when there were dense adhesions and it was believed that excision of the last bit of the tumor might compromise facial nerve function.

Following the craniectomy, cerebrospinal fluid (CSF) drainage from the cerebellomedullary cistern enabled safe retraction of the cerebellar hemisphere. Cerebellar retraction was not started till adequate CSF had been drained and the cerebellum was lax. Peeling of the arachnoid from the posterior surface of the tumor exposed an area sufficient for an initial internal tumoral decompression with the ultrasonic surgical aspirator. Brisk hemorrhage during this step often hampers subsequent arachnoidal dissection, particularly in the lateral or supine–oblique positions, but can be dealt with adequately by gently packing the tumor cavity with gelatin sponge. The principal strategy used at our institution is an arachnoidal dissection proceeding from an inferior to superior and medial to lateral direction. Inferiorly, we dissect the 9th/10th nerves off the inferior pole facilitated by a superior traction on the tumor and by identifying the cochlear nerve usually seen as a thin nerve bundle even in giant tumors adherent to the tumor capsule. Remaining within the plane provided by the cochlear nerve rather than on the arachnoid of the 9th/10th nerves aids in the identification of the facial nerve at its root entry zone (REZ). Intermittent debulking of the tumor is of immense importance making arachnoidal dissection easier and safer.

With the facial nerve identified medially, it is at this stage that the porus acusticus is drilled to remove the
intracanicular tumor and carefully identify the lateral part of the facial and cochlear nerves. Again, preserving the plane of the cochlear nerve is important, not for hearing preservation but for preserving the integrity of the facial nerve. We have found, like others, that the facial nerve courses superiorly from its REZ on the ventral capsule of the tumor, sometimes medially located close to the 5th nerve, but occasionally more laterally. Thus, with the identification of the medial and lateral ends of the facial nerve, a more rapid debulking of the tumor can commence before attempting arachnoid dissection at the superomedial and superolateral regions of the tumor. In the ventral-inferior depths of the field, the 6th nerve is often adherent to the tumor capsule in giant tumors and care is taken to gently displace it by sharp dissection of the arachnoid. Raising the current of the nerve stimulator to 5 and 10 mA at this stage, particularly following substantial debulking of the tumor, might give the surgeon a general idea of the course of the facial nerve, with current passing through 0.5- to 1-cm thickness of the tumor. For a more precise location of the facial nerve, the current is reduced to 0.05 and 0.1 mA. Continuous electromyographic (EMG) recordings from the facial electrodes with an auditory feedback can detect aggressive manipulation of the tumor capsule. This helps in warning the surgeon of the impending facial damage, encouraging him to either be gentler or cease dissection in that region and proceed to another area of the tumor. Arachnoidal dissection proceeds superiorly from the 7th cranial nerve REZ, dissecting the tumor from the brain stem and the 5th nerve, and alternating with a gentler tumor debulking using the ultrasonic aspirator. It is at this point in the surgery that the surgeon might wish to stop and leave behind a part of the tumor should the patient have requested for 100% facial nerve function. Further surgery definitely increases the risk to facial function, as the facial nerve is splayed out over a large area of the tumor. A careful dissection proceeding from the medial to lateral and lateral to medial directions along the facial nerve may prevent anatomical damage to the nerve and effect a radical excision of the tumor.

Hemostasis is secured with oxidized cellulose and is confirmed by asking the anesthesiologist to administer the Valsalva maneuver. Particular attention to hemostasis on the cerebellar surface helps to prevent the development of postoperative hematomas, and should there be a doubt with regard to the presence of a cerebellar contusion, we advocate a generous removal of the involved area rather than mere placement of hemostatic agents. Some surgeons closed the dura using a dural substitute or pericranial graft, while others approximated it and covered it with gelatin sponge.

Postoperative management
Routine postoperative ventilation was not resorted to. Ventilation was performed in patients in whom the surgery was unduly prolonged (e.g., >10 hours) or when the hemostasis was particularly difficult to achieve. These patients were typically extubated the next morning. Tracheostomy was rarely required. All patients received dexamethasone (16 mg/day for 3 days). Prophylactic antibiotics (ceftriaxone or chloramphenicol) were administered only in the first 24 hours after surgery. Tarsorrhaphy was performed for all patients with inadequate eye closure. Oral feeds were started after ensuring that swallowing of liquids was possible without any coughing. A hoarse voice and a poor cough were indications to delay oral feeding. A contrast-enhanced CT scan was done after surgery in all patients at 1 week to determine the extent of tumor removal. Residual tumors were treated with stereotactic radiosurgery if found to be increasing in size at follow-up.

Follow-up
We followed 113 of the 179 (63%) patients, with the follow-up duration ranging from 4 to 72 months (mean, 18.1 ± 12 months).

Statistical analysis
Categorical data are presented using frequency and percentage, and continuous variables are presented as mean and standard deviation. Data were entered into a database (Microsoft Excel) and then analyzed. All statistical analyses were performed using SPSS 11.0 for Windows (IBM Inc.).

Results

Clinical and radiological presentation
The mean age at presentation in Group A was 41.8 ± 13.05 years. This was significantly higher when compared with patients in Group B (≥5 cm), who presented 5–6 years earlier at a mean age of 34.8 ± 12.3 years. Papilledema (66.9% vs. 81.8%) and VI CN dysfunction (2.4% vs. 9.1%; P < 0.05) were significantly more prevalent in Group B [Table 1]. There was no difference in the incidence of hydrocephalus between the two groups, or in the consistency of the tumor [Table 2].

Shunt surgery
Approximately 14% of patients underwent a preoperative shunt in both the groups either in our institute or elsewhere and were then referred here for definitive management of the tumor. Shunts were performed by us in only 20 (11.5%) of 174 patients (excluding the 5 patients who received shunts from elsewhere). Five patients (one in Group A and
four in group B) required a postoperative shunt at varying time periods from 1 to 6 weeks and, in one patient, the shunt inserted preoperatively was removed after surgery.

**Extent of Resection (EOR) and facial nerve outcomes**

Table 3 describes the extent of resection and the facial nerve outcomes [Table 4] in both the groups. There was no difference in the rate of total excision (86.2% vs. 85.4%), anatomical and physiological facial nerve preservation rates between the two groups (approximately 2/3 and 1/3, respectively), and the facial function at discharge. However, at the last follow-up, 65%–75% of patients had either a good (H and B grade 1–3) or a fair (H and B grade 4) outcome.

**Other surgical outcomes**

The overall mortality in the series was 2.2% (4/179). A nasogastric (NG) tube had to be inserted in the postoperative period in 15.5% of patients (27/174; excluding the 5 patients who had a preoperative NG tube in place). Tracheostomy was done in the postoperative period in 4.4% of patients (8/179). Two of these patients had a tracheostomy done preoperatively owing to severe lower cranial nerve dysfunction. Table 5 depicts that while there was no significant difference in the postoperative outcomes between the two groups, the incidence of postoperative CSF rhinorrhea was much higher in Group B (10.9% vs. 2.4%; P < 0.05).
While at the first glance, it might seem that an increase in the tumor diameter by 1 cm (from 4 to 5 cm) is inconsequential, the corresponding volumes tell a different story. A 4-cm spherical tumor would have a volume of $32 \text{ cm}^3$, while in a 5-cm sphere, the volume exponentially increases to $62 \text{ cm}^3$. This substantial increase in the tumor volume within the posterior fossa explains the poorer clinical condition and outcomes seen in the subgroup of patients with tumors larger than 5 cm. With increasing displacement of the eloquent structures, the facial nerve is further splayed out and the brainstem is even more indented, making preservation of function at surgery more difficult. Not only is there an impact on audio-facial morbidity, but also the effect on quality of life cannot be overemphasized.

While there are only a few studies that discuss the outcomes and technical surgical nuances specific to giant VSs (>4 cm in size), to our knowledge, there is no comparison within this group with a larger subset of tumors that are >5 cm in size available in the English literature.

### Discussion

We found that patients with tumors ≥5 cm in size had a shorter duration of symptoms than those with tumors ≥4 cm in size. While the incidence of hydrocephalus was comparable between the two groups, it is likely that raised intracranial pressure was greater in the group with tumors ≥5 cm in size, as evidenced by the increased incidence of papilledema at earlier presentation. Kumar et al., have shown that independent of the tumor volume, elevated cisternal CSF proteins may play an important role in determining the presence of papilledema and visual outcome in giant VSs. The presence of an increased incidence of diplopia due to a unilateral sixth nerve paresis in this group with tumors ≥5 cm in size also may be attributed to the size of the tumor and direct compression of the abducens nerve rather than a generalized raised intracranial pressure, which causes a bilateral lateral rectus paresis.

### Requirement for shunts

There is now sufficient evidence to suggest that hydrocephalus related to giant VSs resolves with the removal of the tumor, obviating the need for a CSF diversion procedure such as

### Table 5: Postoperative complications

<table>
<thead>
<tr>
<th></th>
<th>Group A (4-4.9 cm) N=124 (69.3%)</th>
<th>Group B (≥5 cm) N=55 (30.7%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Postoperative ventilation</td>
<td>12 (8.6)</td>
<td>3 (5.4)</td>
</tr>
<tr>
<td>Reason for postoperative ventilation in 15 patients</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Long surgery</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Postoperative respiratory infection</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Hematoma (tumor bed/EDH)</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>Vocal cord palsy</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Nasogastric (NG) tube</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Preoperative NG tube</td>
<td>2 (1.6)</td>
<td>3 (5.4)</td>
</tr>
<tr>
<td>Discharged with NG tube</td>
<td>20 (16.1)</td>
<td>12 (21.8)</td>
</tr>
<tr>
<td>Data available for 15 of 32 patients who were discharged on Ryle’s tube</td>
<td></td>
<td></td>
</tr>
<tr>
<td>NG tube removed at follow-up (9)</td>
<td>7</td>
<td>2</td>
</tr>
<tr>
<td>NG tube not removed at follow-up (6)</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Tracheostomy (2 of 8 patients had a preoperative tracheostomy)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Postoperative tracheostomy</td>
<td>6 (4.8)</td>
<td>2 (3.6)</td>
</tr>
<tr>
<td>Closed at follow-up</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Data available for 5 of the 8 patients who had postoperative tracheostomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bacterial meningitis</td>
<td>4 (3.2)</td>
<td>2 (3.6)</td>
</tr>
<tr>
<td>Postoperative hematoma</td>
<td>7 (5.6)</td>
<td>1 (1.8)</td>
</tr>
<tr>
<td>Tumor bed</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Extradural</td>
<td>1</td>
<td>0</td>
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<tr>
<td>Spontaneous</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Surgery for hematoma</td>
<td>6/7</td>
<td>0/1</td>
</tr>
<tr>
<td>CSF wound leak*</td>
<td>12 (9.6)</td>
<td>7 (12.7)</td>
</tr>
<tr>
<td>CSF rhinorrhea</td>
<td>3 (2.4)</td>
<td>6 (10.9)*</td>
</tr>
<tr>
<td>Surgery for CSF rhinorrhea</td>
<td>3/3</td>
<td>5/6</td>
</tr>
<tr>
<td>Resolved with LSAD</td>
<td>0/0</td>
<td>1/6</td>
</tr>
<tr>
<td>Mortality</td>
<td>2 (1.6)</td>
<td>2 (3.6)</td>
</tr>
</tbody>
</table>

*All CSF wound leaks stopped with either resuturing or LSAD (lumbar subarchnoid drain)
a shunt or a ventriculostomy.\textsuperscript{[15,16]} In the current series, approximately 14% of patients, irrespective of their size, underwent a shunt to tide over the emergency, as many patients presenting with raised pressure and severe ataxia in the advanced stage of the disease could not be operated on immediately. Currently, we try to operate on the tumor directly, when feasible. However, there was no difference in the requirement of a preoperative shunt between both the groups.

**Operative outcomes**
While some authors prefer to stage surgery for giant tumors to achieve total excision,\textsuperscript{[17,18]} others believe in a single staged procedure.\textsuperscript{[3]} The current trend that seems to be gaining popularity is a near-total excision followed by radiosurgery to improve facial nerve outcomes. Whether this is the optimal form of treatment remains to be proven in long-term follow-up studies. On the other hand, some studies have shown excellent rates of total excision in giant tumors (95%–100%), with very good functional facial nerve outcomes (70%–75%).\textsuperscript{[5,19,20]} While we could achieve a total excision rate of approximately 85% in both the groups, there was no difference in the rate of anatomical and physiological facial nerve preservation rates between the two groups (approximately 2/3 and 1/3, respectively) and the facial function at discharge. However, approximately 65%–75% of patients had a favorable outcome at a long-term follow-up. Most patients who were discharged with an nasogastic (NG) tube had their tube removed within 3–6 months after surgery.

**Surgical nuances**
All surgeons adhered to the surgical principles documented earlier in the text. The salient features of these principles include adequate CSF drainage to obtain a lax cerebellum, avoidance of vigorous retraction of the cerebellum, an inferior to superior progression in tumor resection, early identification of the facial nerve medially, dissection between the identified cochlear nerve and tumor capsule, drilling of the posterior lip of the porus, and continuous facial nerve monitoring. Close monitoring in the postoperative period, especially with respect to breathing and swallowing functions, are essential for a good outcome. Delaying oral feeds in patients with even a mild swallowing dysfunction prevents pneumonia. Protection of the eye in those with significant facial palsy in the postoperative period with liberal use of lateral tarsorrhaphy prevents corneal ulceration and visual loss.

**Postoperative complications**
The incidence of postoperative complications (hematoma, meningitis, CSF leak) was comparable to that described in literature. Interestingly, the incidence of postoperative morbidity was not statistically different between the two groups, except for the occurrence of postoperative CSF rhinorrhea, which was significantly greater in the larger-tumor group (10.9% vs. 2.4%). On reviewing the MR images in these patients, we found that in these cases, the tumor had caused extensive erosion of the internal auditory canal, probably exposing the air cells within the posterior lip of the meatus. The rhinorrhea resolved in all cases after plugging the air cells with fat and glue. This emphasizes the importance of paying special attention to the canal during resection of much larger tumors.

**Limitations**
The limitations of this study are inherent to its retrospective nature. Nonetheless, this is one of the largest series published on giant VSs providing insight into the clinical presentation and immediate postoperative outcomes and comparing them with a subgroup of even larger tumors, an analysis that has not been conducted before.

**Conclusions**
Patients with VSs ≥5 cm in size were younger, with a higher incidence of papilledema and lateral rectus paresis. However, when compared with tumors <5 cm in size, there was no difference in the EOR, facial nerve preservation, and postoperative complications (except CSF rhinorrhea) or mortality. Thus, it is evident from this study that further subclassification of giant tumors is not necessitated. The surgical nuances documented were adhered to by all surgeons operating on patients included in this study. This enabled us to achieve remarkably good outcomes even when these difficult tumors were operated on by multiple surgeons with different levels of expertise.

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**Conflicts of interest**
There are no conflicts of interest.

**References**


