Extent of resection and early postoperative outcomes following removal of cystic vestibular schwannomas: surgical experience over a decade and review of the literature

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Object. Vestibular schwannomas (VSs) are benign tumors of the eighth cranial nerve sheath, representing approximately 6%–8% of all newly diagnosed brain tumors, with an annual incidence of 2000–2500 cases in the US. Although most of these lesions are solid, cystic vestibular schwannomas (CVSs) compose 4%–20% of all VSs and are commonly larger at the time of presentation. The authors present their experience with the operative management of CVSs, including surgical approach, extent of resection, and postoperative facial nerve outcomes. The literature pertaining to clinical and histopathological differences between CVSs and their solid counterparts is reviewed.

Methods. The University of Southern California Department of Neurosurgery database was retrospectively reviewed to identify patients who had undergone resection of a VS between 2000 and 2010. One hundred seventy-nine patients with VS were identified. Patients with CVSs were the subject of the present analysis. Diagnosis of a CVS was made based on MRI findings. Clinical and neuroimaging data, including pre- and postoperative assessments and operative notes, were collected and reviewed.

Results. Twenty-three patients, 14 men (61%) and 9 women (39%), underwent 24 operations for CVSs. These patients composed 12.8% of all cases of VS. Patient ages ranged from 28 to 78 years (mean 55 years), and the mean maximal tumor diameter was 3.6 cm (range 2.0–4.0 cm). Patients most frequently presented with headache, hearing loss, vertigo, and dizziness. Preoperative facial numbness was reported in 44% of patients. Among the 24 cases, 13 were treated with retrosigmoid craniotomy and 11 via a translabyrinthine approach. Complete resection was achieved in 11 patients (48%), subtotal resection (STR) in 8 patients (35%), and near-total resection (NTR) in 4 patients (17%). Facial nerve outcomes were available in all except one case. Good facial nerve outcomes (House-Brackmann [HB] Grades I–III) were achieved in 82% of the patients who had undergone either NTR or STR, as compared with 73% of patients who had undergone gross-total resection (GTR; p > 0.05, Fisher exact test). In comparison, 83% of patients with solid VSs had a good HB grade (p = 0.38, Fisher exact test), although this finding did not reach statistical significance. Complications included wound infection (2 patients), delayed CSF leakage (1 patient), and a delayed temporal encephalocele following a translabyrinthine approach and requiring surgical repair (1 patient).

Conclusions. Cystic vestibular schwannoma represents a clinical and surgical entity separate from its solid counterpart, as demonstrated by its more rapid clinical course and early surgical outcomes. Facial nerve grades may correlate with the degree of tumor resection, trending toward poorer grades with more significant resections. Although GTR is recommended whenever possible, performing an STR when facial nerve preservation is in jeopardy to improve facial nerve outcomes is the preferred strategy at the authors’ institution.

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Key Words • cystic vestibular schwannoma • retrosigmoid approach • translabyrinthine approach • facial nerve

Abbreviations used in this paper: CVS = cystic vestibular schwannoma; GTR = gross-total resection; HB = House-Brackmann; NTR = near-total resection; RS = retrosigmoid; STR = subtotal resection; TL = translabyrinthine; USC = University of Southern California; VS = vestibular schwannoma.

V estibular schwannomas are one of the most common benign intracranial tumors, representing 6%–8% of all newly diagnosed brain tumors, with an approximate annual incidence of 2000–2500 cases in the US. Specifically, they compose approximately 80% of the tumors arising within the cerebellopontine angle. Although the majority of VSs are solid, a small proportion are cystic.

This cystic variety has been reported to account for 5.7%–48% of these tumors, although more recent studies have documented an incidence of 10% among all VSs. However, the accuracy of these estimates is...
debatable given the variety of definitions of what constitutes a CVS. Radiographically, CVSs are best diagnosed on MRI studies, with the presence of fluid-filled hyperintense compartments on T2-weighted sequences and, typically, hypointense compartments on T1-weighted imaging (Fig. 1). Epidermoid and arachnoid cysts, also in the differential diagnosis, can be distinguished from CVSs by the presence of Gd enhancement on MRI, which is generally not noted in the former pathologies.

The cystic variety of tumors can also vary from its solid counterparts in its clinical presentation. Solid tumors have been reported by Selesnick and Johnson to grow at a rate of about 2–6 mm per year. Conversely, cystic tumors can demonstrate a more unpredictable growth pattern, with rapid expansion of the cystic elements. These lesions can distort the brainstem, stretch cranial nerves, and lead to a rapid rise in intracranial pressure, without allowing the brain to gradually compensate for such compressive effects. As a result, patients presenting with CVSs may have a higher incidence and more rapid clinical course of elevated intracranial pressure, papilledema, hydrocephalus, cranial nerve paresis, and facial paresthesias.

Management paradigms for CVSs include clinical observation, radiosurgery, open resection, or a combination of surgery and radiosurgery. When the goal of surgery has been GTR, reported outcomes for facial nerve function have unfortunately been less successful than those for solid VSs, which can be attributed to the adherent nature of these cystic tumors to surrounding structures.

In this paper we review our experience in the surgical treatment of CVSs, including surgical approached used, extent of resection, and early postoperative facial nerve outcomes. We also briefly review the clinical and histopathological differences between CVSs and their solid counterparts.

Methods

A search of the USC Department of Neurosurgery patient database was conducted to identify patients who had undergone craniotomy for the treatment of VS between the years 2000 and 2010. Only patients with CVSs were included in our analysis, and those treated solely with observation or stereotactic radiosurgery were excluded. Patient charts were retrospectively reviewed, including clinical data and notes, operative reports, pathology reports, and neuroimaging results. Data were reviewed in a confidential manner after obtaining approval from the institutional review board at the Keck Hospital of USC and LAC + USC Medical Center, in accordance with the Health Insurance Portability and Accountability Act.

All patients with CVS underwent a rigorous preoperative study panel. Computed tomography imaging data were available in most patients, whereas all patients underwent Gd-enhanced MRI studies. Audiometry testing was also performed in all patients, most importantly in those for whom a translabyrinthine approach was recommended. Gross-total resection was reported in patients who demonstrated no evidence of macroscopic tumor at the time of surgery. In cases in which a small remnant of tumor was left on the facial nerve or brainstem, NTR was reported. Any larger residual tumor was reported as STR.

Surgical approaches used in our patient population consisted of either retrosigmoid or translabyrinthine craniotomy. The choice depended on the presence or absence of serviceable hearing on preoperative testing, as well as the extent of tumor within the internal acoustic canal, regardless of tumor size. Patients were positioned either supine with the head turned 90° or, when neces-
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necessary, in a lateral or park bench position. In the operative suite, a neurophysiologist monitored facial nerve function as well as brainstem auditory evoked responses, somatosensory evoked potentials, and motor evoked potentials. Whenever possible, the Cavitron ultrasonic surgical aspirator (CUSA) was used. Postoperative assessment of facial nerve function was performed using the HB grading system. For this study, we reported only early patient follow-up, including significant postoperative events and complications, early clinical follow-up, and neuroimaging study results. The senior author (S.L.G.) performed all operations.

Results

We identified 179 patients who had undergone resection of a histologically verified VS at USC during a 10-year period (2000–2010). Twenty-three patients (12.8%) had a CVS, as determined by neuroimaging characteristics. Fourteen patients (61%) were men, and 9 (39%) were women, with a mean age of 55 years (range 28–78 years). Sixty-seven percent of the tumors were located on the right, and all patients had single (unilateral) tumors on preoperative imaging, with no history of neurofibromatosis Type 2. The most frequent presenting symptom was diminished hearing in the ipsilateral ear (94%), followed by vertigo (69%), facial sensory disturbances (44%), and headaches (31%; Table 1). All patients were treated primarily at our institution, without any prior radiation or surgical therapy. Magnetic resonance imaging was performed in all patients, and the mean maximal tumor diameter was 3.6 cm (range 2.5–4.0 cm).

Surgical and Facial Nerve Outcomes

The two surgical approaches favored by the senior author (S.L.G.) for CVSs are retrosigmoid and translabyrinthine craniotomy. Twelve patients (52%) underwent a primary retrosigmoid approach and the remainder (48%) underwent a translabyrinthine approach. After presenting with symptomatic tumor progression, 1 patient underwent a retrosigmoid craniotomy 21 months after the translabyrinthine approach. Subtotal resection was achieved in 8 patients (35%), NTR in 4 (17%), and GTR was possible in 11 (48%). Subtotal tumor resection was possible in 42% (5 of 12) of the patients who underwent the retrosigmoid approach and 27% (3 of 11) of those who underwent the translabyrinthine approach. Near-total resection was documented in 17% (2 of 12) of those who underwent the retrosigmoid approach and 18% (2 of 11) of those who underwent the translabyrinthine approach. Lastly, in patients with complete tumor resection, 42% (5 of 12) underwent the retrosigmoid approach, and 55% (6 of 11) the translabyrinthine approach. The most common reason for NTR or STR was tumor adherence to or invasion of surrounding vital structures (facial nerve, brainstem, and so forth) or difficulty in accurately identifying the facial nerve.

Facial nerve outcomes were available in all but 1 patient (this patient underwent STR via a retrosigmoid approach). Postoperative facial nerve outcomes were good (HB Grade I–III) in 77% (17 of 22) of the patients. Among them, we noted HB Grade I in 65% (11 of 17), Grade II in 12% (2 of 17), and Grade III in 24% (4 of 17; Fig. 2). A good HB grade was achieved in 86% (6 of 7) of the patients with STR, 75% (3 of 4) of those with NTR, and 73% (8 of 11) of those with GTR (p > 0.05). A poor HB grade (Grades IV–VI) was evident in 23% of the patients overall: A poor HB grade (IV–VI) facial palsy developed in 14% (1 of 7) of patients with STR, 25% (1 of 4) of those with NTR, and 27% (3 of 11) of those with GTR. Good HB outcomes were obtained in 82% (9 of 11) of patients who underwent either NTR or STR, as compared with 73% of those who underwent GTR (p > 0.05, Fisher exact test). Anatomical preservation of the facial nerve was noted in all 23 patients.

As a comparison, in patients with solid VSs, 83% had a good HB grade (p = 0.38, Fisher exact test). A good HB outcome was obtained in 89% (16 of 18) of those who underwent STR for a solid VS and 83% (114 of 138) of those who underwent GTR of a solid VS. These results were not statistically significant, as compared with those in patients with CVSs.

Patient Complications

No patients undergoing resection of a CVS died. Wound infections without meningitis developed in 2 patients postoperatively, requiring wound revision. One of these patients, who initially underwent a translabyrinthine approach followed by a retrosigmoid approach 21 months later, experienced delayed wound breakdown. One patient presented with delayed CSF leakage 2 years after a retrosigmoid approach, and the leak was surgically

<table>
<thead>
<tr>
<th>TABLE 1: Neurological symptoms on presentation with CVS</th>
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<tr>
<td>Symptom</td>
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<td>--------------------------</td>
</tr>
<tr>
<td>hearing loss</td>
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<tr>
<td>vertigo/dizziness</td>
</tr>
<tr>
<td>facial sensory disturbance</td>
</tr>
<tr>
<td>headaches</td>
</tr>
<tr>
<td>facial nerve symptoms or weakness</td>
</tr>
<tr>
<td>tinnitus</td>
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</tbody>
</table>

Fig. 2. Bar graph depicting HB facial nerve function outcomes in patients who underwent craniotomy for resection of CVSs (black bars) and solid VSs (gray bars). The y axis represents the percentage of patients.
repaired. Another patient presented with seizures 2 years after resection of a CVS via the translabyrinthine approach. Follow-up MRI demonstrated what appeared to be a temporal lobe encephalocele in the translabyrinthine defect. The patient had complete resolution of seizures following surgical repair of this lesion.

Discussion

Vestibular schwannomas are benign tumors believed to arise from Schwann cells of the eighth cranial nerve. The annual incidence in the US is approximately 10–15 cases/million people.10,25 Although VSs are generally considered slow-growing neoplasms, the cystic subtype can grow quite rapidly. As a result, the presentation for CVS can differ from that for solid VS, including a shorter duration of symptoms prior to presentation, increased risk of cranial nerve palsy, and increased rates of hydrocephalus.

Previous reports have suggested that CVSs compose 5.7%–48% of all VSs. More recent studies have favored a proportion closer to 10%.6,11,12,23 Our series corroborates the findings of more recent studies, demonstrating an incidence of 12.8% among all patients with VS (Table 2). However, it is important to note that our data are based on neuroimaging findings alone. In a recent study by Fundová et al.,6 in which 2 additional criteria were used for the diagnosis of CVS (intraoperative identification of cystic elements and histopathological staining for S100 protein positivity in addition to radiographic data), an incidence of only 5.7% was reported.

The precise etiology of the cystic component of these tumors remains unknown, although various mechanisms for cystic formation have been proposed. Although not mutually exclusive, these mechanisms have included tumor growth with subsequent central necrosis, coalescence of microcysts within the Antoni B environment, and/or repeated hemorrhage within the tumor.2,18 Cyst enlargement can subsequently occur because of an osmotic gradient resulting from serum proteins or the production of mucinous material within the cyst.2,23 In a more recent report by Moon et al.,17 matrix metalloproteinases—proteolytic enzymes found naturally during embryogenesis and tissue remodeling—were identified within the cyst fluid and walls of CVSs. These authors believe that matrix metalloproteinase-2 may contribute to cyst formation as well as tumor adhesion to the facial nerve by promoting tumor expansion and growth or by stimulating proteolytic degradation at the tumor-nerve interface. Future studies analyzing the genomic and epigenetic substrates for a CVS phenotype may provide more information regarding the etiology of this cystic formation.

Cystic vestibular schwannomas have been associated with more rapid tumor growth, a shorter duration of symptoms, and increased involvement of the facial nerve.5,6 As evident in our cohort of patients, the symptoms most commonly found in patients presenting with CVS include hearing loss, headache, cerebellar signs, trigeminal nerve involvement, and facial nerve involvement. Other signs, such as tinnitus, lower cranial nerve paresis, diplopia, and visual symptoms or loss, can also be seen.23

Surgical outcomes after craniotomy for CVS have been reported to be worse than those for solid VSs, specifically when evaluating facial nerve function. In the current study, worse facial nerve outcomes were associated with GTR of a CVS rather than a solid VS, although these results did not reach statistical significance. A previous report by Fundová et al.6 demonstrated a poor HB grade (IV–VI) in 66% of patients at 1 year or more of follow-up. In the report by Sinha and Sharma,23 facial nerve preservation rates were 67.9% and 82.7% in the CVS and solid VS groups, respectively. A good HB grade (I–III) was noted postoperatively in 67.9% of patients with CVS. In a more recent report by Piccirillo et al.,20 81% of patients with CVSs who underwent resection had a good HB grade. All of these studies demonstrate improved facial nerve outcomes in solid VS cases as compared with outcomes in CVS cases. Our results correspond with previous findings: 77% of patients with CVSs who underwent treatment demonstrated an HB grade between I and III, as compared with 83% in solid VS cases.

The role of stereotactic radiosurgery has yet to be elucidated in the treatment of CVS. In a report by Pendell et al.,19 radiosurgery outcomes were suboptimal, with 3 of 6 cystic tumors demonstrating rapid and significant cystic expansion requiring urgent surgery for neurological decline. Ganslandt et al.7 described an intratumoral hemorrhage leading to death in a patient with CVS 15 months after treatment with stereotactic radiosurgery. Delsanti and Régis4 described their experience with stereotactic radiosurgery in the treatment of 54 CVS cases. Their failure rate, defined as the need for a second procedure, was 6.4%, almost 3-fold greater than that reported for solid VS.12 In our series, only 1 patient was treated with radiosurgery (5040 cGy in the form of intensity-modulated radiation therapy). As can be seen in Fig. 3, despite 2 resections and adjuvant radiation therapy, the CVS recurred after each intervention, with ongoing cyst formation. Surprisingly, however, almost 2 years after external radiotherapy, the cystic and solid components of the tumor demonstrated a noticeable decrement in size. Cystic VSs with multiple recurrences, in particular, pose a significant challenge in achieving a surgical cure or tumor control.

TABLE 2: Literature survey of the incidence of CVS

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Total No. of VSs</th>
<th>No. of CVSs (% of total)</th>
</tr>
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<tbody>
<tr>
<td>Kendall &amp; Symon, 1977</td>
<td>31</td>
<td>3 (9.7)</td>
</tr>
<tr>
<td>Robbins &amp; Marshall, 1978</td>
<td>39</td>
<td>8 (20.5)</td>
</tr>
<tr>
<td>Wallace et al., 1993</td>
<td>35</td>
<td>7 (20)</td>
</tr>
<tr>
<td>Tali et al., 1993</td>
<td>80</td>
<td>15 (18.8)</td>
</tr>
<tr>
<td>Charabi et al., 1994</td>
<td>571</td>
<td>23 (4)</td>
</tr>
<tr>
<td>Jeng et al., 1995</td>
<td>27</td>
<td>13 (48)</td>
</tr>
<tr>
<td>Pendell et al., 1996</td>
<td>148</td>
<td>9 (6.1)</td>
</tr>
<tr>
<td>Fundová et al., 2000</td>
<td>773</td>
<td>44 (5.7)</td>
</tr>
<tr>
<td>Sinha &amp; Sharma, 2008</td>
<td>284</td>
<td>58 (20.4)</td>
</tr>
<tr>
<td>Piccirillo et al., 2009</td>
<td>1416</td>
<td>96 (6.8)</td>
</tr>
<tr>
<td>present study</td>
<td>179</td>
<td>23 (12.8)</td>
</tr>
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Conclusions

Cystic vestibular schwannomas represent an entity separate from their solid counterparts, as demonstrated by their greater likelihood for facial numbness and hydrocephalus at presentation, rapid clinical course, adherence to vital neurological structures, and early surgical outcomes. Early facial nerve function appears to correlate with the degree of tumor resection, with poorer grades generally noted with more radical resections. We recommend GTR whenever possible, performing STR when facial nerve preservation is in jeopardy to improve facial nerve outcome and limit complications. Although anecdotal, our experience suggests that “once a CVS, always a CVS,” often making the treatment of recurrent CVS a more substantial challenge.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Yashar, Zada, Giannotta. Acquisition of data: Yashar, Harris, Giannotta. Analysis and interpretation of data: Yashar, Harris, Giannotta. Drafting the article: Yashar, Zada, Harris. Critically revising the article: Yashar, Zada, Giannotta. Reviewed submitted version of manuscript: Yashar, Zada, Giannotta. Approved the final version of the manuscript on behalf of all authors: Yashar. Statistical analysis: Yashar. Administrative/technical/material support: Yashar, Zada, Giannotta. Study supervision: Giannotta.

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Fig. 3. Images obtained in a 66-year-old woman presenting with hearing loss, right facial sensory disturbance, and vertigo. Preoperative T1-weighted contrast-enhanced MR image (A) demonstrating a right VS with multiple small cysts and a single large medial cyst adjacent to the brainstem. Axial T2-weighted MR image (B) revealing the classic appearance of CVS with hyperintense signal within the cyst. Postoperative MR image (C) obtained after a translabyrinthine approach, showing residual tumor after STR. Axial MR image (D) obtained 1 year after the image featured in panel B, demonstrating interval development of a large medial cyst with significant compression of the pons and cerebellum. Postoperative axial MR image (E) obtained immediately after a second craniotomy (retrosigmoid approach), demonstrating significant debulking of the tumor, brainstem decompression, and cyst resection. Follow-up MR image (F) obtained 8 months later, showing tumor recurrence along with 2 small medial cysts, which prompted adjuvant external radiation treatment with 5040 cGy. Axial MR image (G) obtained 2 years after the completion of radiation, showing a mild decrease in the size of the 2 small cysts and the residual tumor.

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