Surgical treatment of large vestibular schwannomas (stages III and IV)

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KEYWORDS
Large vestibular schwannoma; Acoustic neurinomas; Residual vestibular schwannoma; Facial nerve; Surgical results

Summary
Objective: Study the results of surgical treatment of large vestibular schwannomas.
Material and methods: Between January 1995 and December 2005, 87 stage III and IV unilateral vestibular schwannomas (Koos classification) were operated.
Results: The approach used was for the most part translabyrinthine. Nine patients were operated in two phases. Tumor exeresis was total in 79% of the cases, nearly total in 17%, and subtotal in 3.6%. Good facial function was preserved in 63% of the patients. Mortality was zero. Tumor control after a mean follow-up of 45 months was 86%.
Conclusions: Surgery for large vestibular schwannomas should have a 0% mortality rate and low morbidity. Otoneurosurgical collaboration, with a preference for the translabyrinthine approach, with surgery undertaken in several phases if need be, provides maximum safety and good functional results as well as an acceptable residual tumor rate.
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Introduction
Vestibular schwannomas can evolve slowly into large tumors. Symptoms generally begin with strictly otological symptoms that can be ignored by the patient. Then, as they enlarge, the tumor reaches the central nervous system, progressively showing cranial nerve involvement, cerebellar or brainstem problems, and obstructive internal hydrocephalus. Large vestibular schwannomas therefore affect the vital prognosis and their treatment cannot be conservative because radiotherapy may induce internal hydrencephalus. Surgical removal is therefore the best adapted treatment and is possible even in the oldest patients because of the low morbidity of the approaches used.

However, even if the choice of surgery seems easy to make, it exposes the patient to vital risks, serious complications, and functional sequelae that are significantly greater for large tumors. Surgical management of patients with large vestibular schwannomas should therefore provide maximum safety for the patient, in both vital and functional terms. Preserving facial motor function remains the priority functional consideration, now that it is recognized that the chances of pre-
Figure 1 Koos classification. Stage I: tumor confined to internal auditory canal, diameter 1–10 mm; stage II: tumor extension to external auditory canal, less than 20 mm; stage III: tumor extension to external auditory canal, less than 30 mm or touching the brainstem without causing compression; stage IV: tumor extension to external auditory canal, greater than 30 mm or causing brainstem compression (reproduced from Koos et al. [1]).

serving hearing in large vestibular schwannomas are low.

Patients and methods

Of the 1200 tumors of the cerebellopontine angle operated by the otoneurosurgical team in the past 30 years in our hospital, we have studied the large vestibular schwannomas treated with first-line surgery between January 1995 and December 2005. Stage III and IV tumors according to the Koos [1] morphological classification were selected, i.e., those touching the brainstem (Fig. 1). This classification has the advantage of better assessing the relation between tumor size and posterior cerebral fossa size. The cases of type II neurofibromatosis were excluded as were all incomplete clinical files. A total of 87 patients were finally retained for this retrospective study. Patient follow-up was closed on 1 January 2009 so that the last patients included would have 3 years of follow-up.

Functional work-up

Hearing was classified according to the American Academy of Otolaryngology Head and Neck Surgery (AAO-HNS) guidelines [2]. Classes A and B define serviceable hearing.

Facial motor function was assessed using the House and Brackmann (HB) classification [3]. Good results in facial motor function correspond to grades 1 and 2.

Surgical technique

All patients were operated on by the same otoneurosurgical team with systematic intraoperative monitoring of the facial nerve using a four-channel NIM™ 2.0 (Medtronic) and in certain cases, monitoring of the trigeminal nerve and the brainstem with somesthetic evoked potentials. A robotized microscope-based neuronavigation system (Zeiss MKM™ system and Medtronic’s Surgiscope™) was used for very large tumors or during certain revisions for evolving tumor residue.

Tumors were excised beginning with hollowing the interior of the tumor using a Cavitron® ultrasonic aspirator. Then, the tumor shell was dissected in the subarachnoid plane so as to leave the facial nerve protected by a thin veil and to preserve its vascularization. For the stage IV tumors, a two-phase surgical resection strategy was always discussed with the patient. Either the surgical strategy was defined immediately, aiming to reduce the tumor volume during the first phase to remove the compression and edema from the brainstem, or this decision was made intraoperatively in case of dissection problems such as bleeding, adherences, bradycardia, or abnormal somesthetic evoked potentials. Resection was completed in the second phase, in better conditions.

The quality of tumor resection was evaluated by the operator according to the description provided by Bloch et al. [4]. Although eminently subjective, resections were qualified as nearly total if they left only a fragment of the tumor shell and subtotal if the resection left a larger volume in place.

The approaches used were the translabyrinthine, retrosigmoid, and transotic approaches.

Radiographic work-up

The preoperative workup systematically included a CT scan of the petrous part of the temporal bone and magnetic resonance imaging (MRI) including T1-weighted sequences with and without gadolinium injection and T2-weighted sequences. The tumor volume was calculated on the most recent MRIs. All patients had an MRI between 1 and 3 months after surgery and then annual follow-up during the first 5 years.

Statistical analysis

The qualitative values were compared using the Chi² test or the Fisher test of probabilities for small numbers of subjects. The quantitative variables were compared using the Student t-test. The significance threshold was set at 0.05.

Results

The mean age of the 87 patients operated was 50 years (range: 20–77 years). There were 36 stage III tumors and 51 stage IV, with eight giant tumors that extended beyond the midline (15%).

The time from the first clinical signs to diagnosis was a mean 36 months (range, 1–135). Thirty-one patients (36%) were diagnosed early (delay < 12 months), mainly for stage III as opposed to stage IV (57% versus 30%; p = 0.040). Late diagnoses (>5 years) accounted for another 20% of the patients, with no significant difference between the tumor stages (p = 0.807).
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### Table 1
Final facial function in relation to tumor stage (before and after any surgical revision for recurrent tumor).

<table>
<thead>
<tr>
<th>House and Brackmann grade</th>
<th>Number, stage III</th>
<th>Number, stage IV</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>23 (72%)</td>
<td>14 (27%)</td>
<td>37 (45%)</td>
</tr>
<tr>
<td>II</td>
<td>4 (13%)</td>
<td>11 (22%)</td>
<td>15 (18%)</td>
</tr>
<tr>
<td>III</td>
<td>2 (6%)</td>
<td>10 (20%)</td>
<td>12 (14%)</td>
</tr>
<tr>
<td>IV</td>
<td>1 (3%)</td>
<td>10 (20%)</td>
<td>11 (13%)</td>
</tr>
<tr>
<td>V</td>
<td>1 (3%)</td>
<td>2 (4%)</td>
<td>3 (4%)</td>
</tr>
<tr>
<td>VI</td>
<td>1 (3%)</td>
<td>4 (8%)</td>
<td>5 (6%)</td>
</tr>
</tbody>
</table>

### Preoperative symptoms

During the preoperative clinical workup, 93% of the patients reported hearing impairment, 56% tinnitus, 43% balance impairment or dizziness, 10% headaches, 7% neuralgia or facial hypesthesia.

In 23% of the cases, the clinical examination identified a vestibular abnormality, in 7% cerebellar problems, in 13% hypesthesia of the trigeminal nerve, in 7% facial paresis. Five patients had intracranial hypertension, one of whom also presented contralateral hemiparesis.

These neurological anomalies were finally present in 26% of the cases, so that evolved and compressive tumor was suspected based on the clinical examination. All these patients indeed had a stage IV tumor.

On the functional level, half the patients had serviceable hearing (51%).

### Surgical procedure

The five patients with a stage IV tumor who presented a dangerous clinical picture with intracranial hypertension were first fitted with a ventriculoperitoneal shunt a mean 12 days before the intervention. Five other patients presented an edematous aspect of the brainstem on MRI surrounding the tumor. They were operated after short-term corticosteroid therapy. The approaches used were translabyrinthine in 80 cases, retrosigmoid in six cases — for tumors with preservation of serviceable hearing that did not invade the fundus of the internal auditory canal — and transotic in one case for a very anterior tumor in the cerebellopontine angle. Nine stage IV tumors were operated in two phases. The second surgical phase took place a mean 6 months after the first phase (range, 77 days to 17 months).

At the end of the surgical procedure (in one or two phases), resection was total in 69 cases (79%), nearly total in 15 cases (17%), and subtotal in three cases (3.6%).

### Facial function

The facial nerve was anatomically preserved with certainty in 83% of the patients (73 patients), partially damaged in three cases of adherent tumors, and impossible to preserve in one patient. In 10 cases, the facial nerve was not formally recognizable along its entire length. It was either masked by the arachnoid veil left in place or there was a zone in front of the internal auditory meatus that was difficult to control.

Immediate postoperative facial motor function was analyzed in 81 patients. Good facial function (grade I or II) was found in 48% of the patients (n = 39): 70% for stage III tumors and 33% for stage IV. Of the 42 cases of facial paralysis, we observed six cases with secondary onset (6.8%) on average on Day 5. These cases all regressed completely in less than 2 months.

The facial result at a later date and at the end of the initial surgical procedure (in one or two phases), analyzable in 83 patients, showed 63% good results: 84% for stage III and 49% for stage IV (Table 1). Taking into account the surgical revisions for recurrent tumors, the final result, analyzed at the last follow-up for each patient and at the end of a mean follow-up of 45 months, was equivalent: 60% good results: 84% for stage III tumors and 45% for stage IV (p = 0.75).

Identification of the predictive parameters of the final facial result showed that tumor stage played an important role (p = 0.001), as did good immediate facial function (p < 0.001), tumor hemorrhage (p = 0.043), and whether the facial nerve could be stimulated at the end of the intervention (p < 0.001). On the other hand, age, sex, the duration of symptoms, the presence of trigeminal involvement, the level of hearing impairment, vestibular involvement, and the quality of the resection had no influence.

### Complications and mortality

Mortality related to the surgery was zero. Twenty patients (23%) presented potentially severe complications (Table 2),

<table>
<thead>
<tr>
<th>Major complications</th>
<th>Number (%)</th>
</tr>
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<tbody>
<tr>
<td>Cerebrospinal fluid leak</td>
<td>11 (12.6)</td>
</tr>
<tr>
<td>Meningitis</td>
<td>3 (2.8)</td>
</tr>
<tr>
<td>Hematoma of posterior fossa</td>
<td>1 (1.1)</td>
</tr>
<tr>
<td>Ischemic stroke</td>
<td>1 (1.1)</td>
</tr>
<tr>
<td>Mixed nerve impairment</td>
<td>4 (4.6)</td>
</tr>
<tr>
<td>Death, hydrocephalus, oculomotor impairment, gaseous embolism</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Total</td>
<td>20 (22.9)</td>
</tr>
</tbody>
</table>
three of which were particularly serious: a hematoma of the posterior fossa requiring emergency surgery at the 12th postoperative hour, one case of ischemic stroke, and one case of severe hypoxemia pneumopathy secondary to swallowing impairment after involvement of the mixed nerves. Finally, all these patients were cured with no sequelae.

**Residual tumors**

Other than the intraoperative evaluation by the surgeon, tumor residues were systematically searched for on postoperative MRI; 26 residual tumors were identified and five contrast-enhanced linear images were attributed to inflammatory tissue or arachnoid folds. During the follow-up, two other residual tumor images appeared secondarily at 1 and 3 years, bringing the total number of residual tumors to 28 (32%). Of the 15 nearly total excisions, in two cases, no residual tumor was found on imaging. Conversely, of the 69 total resections, 12 presented an image of a residual tumor (17%).

Of the 28 residual tumors monitored, seven shrank or disappeared (25%) and nine remained stable for a mean 4-year follow-up period (32%). Finally, 12 evolved (43%), becoming a tumor recurrence in 13.8% of the patients in the series (86.2% tumor control). These recurrent tumors corresponded to three subtotal resections (100%), six near-total resections (40%), and three resections evaluated as total (4.3%).

These 12 tumor recurrences required surgery in 10 patients and gamma-knife radiosurgery in another. The last patient was lost to follow-up after 3 years of monitoring. The recurrences were reoperated after a mean delay of 5 years and 5 months after the initial excision (range: 2—9.7 years).

One of the patients has not yet achieved complete tumor control after several interventions. At the age of 20 years, he presented a very large tumor that was partially resected. Then, he was lost to follow-up for 9 years. When he was seen again, the residual tumor measured 88 cm³. It was progressively reduced in three new interventions over 16 months, which was hindered by the calcified character of the tumor. Unfortunately, the facial nerve, which had remained functional, was damaged during the last attempt to resect the large residue in front of the brainstem (Fig. 2).

**Discussion**

**Surgical safety**

All causes combined, mortality rates for vestibular schwannoma surgery have dropped substantially over time, with tremendous progress made by surgical teams, and now are approximately 1%. Large tumors have a larger surface in contact with the brainstem, the cerebellum, and the cranial nerves and blood vessels. They expose the patient to greater risk. Death results for the most part from hemorrhagic complications (35% of cases) [5].

For these large tumors where the chances of preserving serviceable hearing are minimal [6], the translabyrinthine approach is the choice approach because it provides maximum safety because of the direct approach to the cerebellopontine angle. This approach can be particularly useful in cases of cerebellopontine angle hematoma, which requires emergency revision surgery, in contrast to the retrolabyrinthine approach, where the cerebellum pushed back by the hematoma can become an obstacle. The translabyrinthine approach, which takes longer and is more delicate, is improved advantageously by otoneurosurgical collaboration. In our series, with 0% mortality, rapid surgical revision of the only case of cerebellopontine angle hematoma saved the patient’s life with no neurological after-effects.

**Step-by-step exeresis**

In our experience, resection of large tumors in several interventions can also reduce operative morbidity. This strategy was adopted for nine patients (10%). In only three cases, it was decided during surgery, with a highly hemorrhagic tumor, an adhering tumor, and a case where the dissection in contact with the brainstem was responsible for severe bradycardia.

During the second intervention, the dissection problems were reduced in three cases, identical in four cases, and
increased in two cases. In five patients who presented an edema of the cerebellum, observed initially, the second surgery was not disturbed by the edema. Comey et al., who operated 83 large vestibular schwannomas in two or three phases, showed that sequential resection did not increase morbidity and made it possible to reduce bleeding and adherences during the interventions [7]. Patni and Kartush made the same observations in their series of 34 tumors [8]. Their technique is very distinctive. It reduces tumor volume during the first intervention via the retrosigmoid approach without reaming the internal acoustic meatus. Then, the excision is continued in a second intervention via the translabyrinthine approach to dissect the facial nerve in intact tissues in the internal auditory canal. They recommend undertaking the second phase after 4 months so that the cerebellar edema will have resorbed, and before 6 months to prevent tumor progression. They also estimate that the facial nerve is the most robust during the second phase, and the anatomy of the region nearly the norm. Their observations indicate that the facial nerve is the most robust during the second phase and the anatomy of the region nearly the norm. Their patients found the second phase less disabling.

**Facial function**

Since mortality in vestibular schwannoma treatment has become exceptional, preservation of facial nerve functions has become the priority for patients. Unfortunately, facial function outcome is less favorable for large tumors [5,9]. It must be emphasized that even early diagnosis of all vestibular schwannomas would not prevent the discovery of tumors that have already evolved, because their symptoms can appear late. As in 36% of the cases in our series, the tumor was diagnosed less than 1 year after the beginning of functional warning signs.

Table 3 reports the results from the literature of surgical series of large vestibular schwannomas [9–16]. A mean good facial result of 54% (range, 44–94%) was observed. This discrepancy in the results can be explained by the absence of uniformity in the definition of “large tumor” and by the variability in tumor measurement techniques, which sometimes include the internal auditory canal portion. In our series, we obtained a good facial function rate, 63%, similar to the results of other teams, in particular the series reported by Deveze et al. [15], who also used the Koos tumor classification. For our patients who presented a recurring tumor, facial function was worsened only slightly by surgical revision of the residue. These revisions involved only patients initially presenting a stage IV tumor. The rate of good facial function decreased from 49 to 45% after surgical revision during the follow-up period. The search for predictive factors of facial motor function in our study showed the highly significant influence of tumor stage as defined by the Koos classification (p = 0.001).

A hemorrhagic tumor was also a risk factor for facial impairment (64% poor facial function versus 33%; p = 0.043). Untimely bleeding makes the facial nerve and the arachnoid plane more difficult to visualize and increases the risk of mechanical (tamponade, aspiration), thermal, or ischemic lesions during coagulation in the immediate vicinity. To prevent injurious dissection of the facial nerve, particularly with adherent tumors, the dissection must occasionally be interrupted and a small tumor fragment left, particularly with benign tumors, threatening only in their large size. In an older patient, a small residue will rarely pose subsequent problems. In a young subject, the risk of having to return to surgery for tumor recurrence is certainly greater, but the absence of facial paralysis is a clear advantage. Even if our results show no correlation between non total resection and a better postoperative facial result (p = 0.475), it is probable that overzealousness in difficult dissections would have worsened the functional result. In a study of 116 vestibular schwannomas larger than 21 mm, Seol et al. showed a significantly better facial result in cases of incomplete resection [17]. Many authors prefer conservative surgery to preserve the nerve structures [8,17]. Analysis of the literature shows that a mean 23% of stage III and IV vestibular schwannoma removals are incomplete, as in our study (21%) [11–14,16,17].

**Tumor recurrence**

To preserve the nerve structures, removal of large vestibular schwannomas sometimes leaves a small tumor residue, which systematically requires MRI follow-up so that it can be
objectively assessed [18]. The distinction between artifact and authentic residue is not always simple. Although it is recognized that linear images generally correspond to scar tissue or arachnoid folds, only prolonged MRI follow-up can diagnose residual tumors [19]. As has been observed herein, it is surprising to find no residual tumor images after certain nearly total excises. Three explanations are possible: the surgeon may have been pessimistic, the follow-up MRI was not of sufficiently high quality, or small, poorly vascularized tumor residues disappear rapidly. This lack of vascularization can also explain the involution or the stability of authentic residues. We observed that only 43% of the residues evolved. These recurrences involved 14% of the patients with a mean follow-up of 45 months. Their incidence in the literature is difficult to evaluate because of the heterogeneity of the data, the lack of precision between residual and recurrent tumors, and the short time period between surgery and radiological follow-up. This follow-up must be prolonged, which may run the risk of losing patients to follow-up and thereby distort the occurrence rate observed. Moreover, this rate is an important parameter in assessing the facial function results. Following up residual tumors in a series of large vestibular schwannomas over 4 years, Godefroy et al. observed results that were very similar to ours, with 46% residual tumors and 6% recurrent tumors [20]. Only the observation of the recurrent tumors is a therapeutic problem. Freeman et al. observed 60% evolving residues [21]. Thomassin et al., with a long-term follow-up of 137 patients, observed a peak of tumor recurrence at 8 years. They therefore recommend following up patients for at least 8 years and longer in young patients because of one case of recurrence discovered 20 years later [22]. We recommend follow-up MRI in the first 2 months after surgery, then at 1, 3, and 5 years, then as needed depending on the presence of tumor residue. At 10 years, with no residue, it is suitable to stop follow-up.

Managing recurrences consists either in abstention and surveillance, possible in elderly subjects, or microsurgical revision or radiosurgery. Our therapeutic policy has mainly been surgical. This remedial treatment exposes the patient again to risks of neurological sequelae. This risk is classically higher than during the initial treatment [21,23]; however, in our series, seven patients out of 10 preserved good facial function after surgical revision.

Conclusion

Treatment of large vestibular schwannomas is surgical, even if the tumor size is the main risk factor for cranial nerve lesions. In this case, we accept the compromise between preservation of facial nerve function and at least good tumor control, particularly since revisions have altered this function only slightly. This surgery should be performed by trained surgical teams whose experience guarantees a low morbidity rate. The mortality rate should tend toward zero and all the operative safety measures should be taken to ensure this outcome. Interventions with a double onneurrosurgical team, preferring the translabyrinthine approach with excision in several phases if necessary, provides a maximum chance of good results in terms of facial function and an acceptable rate of residual tumor.

Conflict of interest

None.

References


