Geniculate Ganglion Tumors: Clinical Presentation and Surgical Results

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Abstract

Objective. Facial nerve tumors are rare lesions mostly located in the geniculate ganglion. This study aims to compare those tumors limited to the geniculate ganglion in terms of clinical features and postoperative outcomes.

Study Design. Case series with chart review.

Settings. University tertiary reference center.

Subjects and Methods. Medical charts were reviewed for 17 patients who had surgery for geniculate ganglion tumor removal (10 hemangiomas, 6 schwannomas, 1 meningioma). Hemangiomas and schwannomas were compared for preoperative facial nerve function, hearing, tumor size, and postoperative outcomes.

Results. Facial palsy was observed in all cases. Regarding the preoperative facial nerve function, severe facial palsy (House-Brackmann grades V and VI) was present in 70% of cases for hemangiomas and for no case of schwannoma (P = .01), although hemangiomas were significantly smaller tumors (P = .01). Hearing loss was observed in 4 cases (23.5%) and was related to tumor volume (P < .0001). A complete excision was achieved in all cases, and a facial nerve graft was performed immediately after interruption in 16 patients (94%). Postoperative facial nerve function was improved or stabilized in 94% of cases. A preoperative House-Brackmann grade VI was shown as a negative factor for postoperative facial nerve function.

Conclusions. Differences in clinical presentations could help in establishing the good therapeutic option depending on the tumor type. Surgery, when indicated, is safe and effective, and postoperative outcomes are not related to tumor type.

Keywords

geniculate ganglion, hemangioma, schwannoma, meningioma, facial nerve graft

Facial nerve (FN) tumors are rare lesions of the petrous bone. They include schwannomas, hemangiomas, and meningiomas. Schwannomas represent 0.8% of petrous bone tumors. They are benign, encapsulated, slow-growing lesions arising from the Schwann cells and can involve any of the different segments of the FN, with the geniculate ganglion (GG) being the most frequent segment involved. FN hemangiomas encompass 0.7% of petrous bone tumors, and they have been reclassified in the group of vascular malformations. As schwannomas, hemangiomas are mainly located in the GG. FN meningiomas are extremely rare, with few cases reported in the literature. They arise from arachnoid cells accompanying the FN during its embryonic formation. The GG and the internal auditory canal are the 2 preferential locations of petrous bone meningiomas.

When the tumor is limited to the GG, the most common symptom is facial paralysis. Even if the radiologic features of these lesions (magnetic resonance imaging [MRI] and computed tomography [CT] scan) might help in the differential diagnosis, no study has analyzed differences in clinical presentation among the tumor types that could help in establishing a correct preoperative diagnosis. This is essential to define the correct management of these tumors, given the increasing number of patients affected by FN schwannoma who are treated with stereotaxic radiosurgery.

Thus, the aim of this study was to compare the different tumor types of GG tumors in regard to their preoperative clinical features and postoperative results.

Methods

Medical records were retrospectively reviewed for patients who had surgery for an intrapetrous FN tumor from 1988 to 2013 in a tertiary referral center. All patients gave their informed consent.

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for the use of their clinical data, and the ethics committee of Pitie-Salpetriere hospital approved the study. All the patients were operated on and evaluated by the same senior surgeon (O.S.) for pre- and postoperative features, because he was the only physician present for the entire duration of the inclusion period and the most experienced one for FN tumor cases.

Inclusion criteria were an isolated GG tumor on the basis of imaging and postoperative histologic examination. Tumors that were located in another segment of the FN or spreading from the GG to another segment were excluded. Nonprimary FN tumors involving the petrous bone with extension to GG (metastasis, cholesteatomas) were also excluded. Patients with a diagnosis of type II neurofibromatosis were excluded because of the risk of multiple lesions involving the FN. The tumor type was confirmed by histologic examination after tumor excision. Frozen sections of the FN at proximal (labyrinthine) and distal (tympanic) segments of the FN were performed in all but 1 patient, who underwent tumor excision without interruption of the FN.

Preoperative Assessment

The data at the first consultation included demographic information, presenting symptoms, facial function according to the House and Brackman (HB) scale,\textsuperscript{10} mean pure-tone audiometry (PTA; mean of 500, 1000, 2000, and 3000 Hz) in air and bone conduction with headphones, hearing classification according to American Academy of Otolaryngology—Head and Neck Surgery Foundation, and vestibular function evaluated by a caloric test. A vestibular impairment was defined as a lateralization and a directional preponderance >25% as calculated by Jongkee’s formula. Imaging (CT scan and MRI in T1-WI, T2-WI, and T1 with contrast) was available for all patients and was analyzed for tumor size on the postcontrast T1-WI sequences and for the presence of cochlear or labyrinthine fistula on CT scan and T2-WI.

Peri- and Postoperative Data

Perioperative data included surgical approach and, in case of interruption of the FN, the type of reconstruction of the nerve. Postoperative data included complications, FN function at 12 months and at the last consultation, auditory outcomes, and recurrences detected by MRI studies performed every year after surgery for 3 years.

Statistical Analysis

Results are presented as mean ± SD. Hemangiomas and schwannomas were compared with Fisher tests for qualitative data and Wilcoxon, Kruskal-Wallis, and Pearson tests for quantitative data. Statistical tests were performed with R (version 3.2.3). Differences were considered statistically significant when $P < .05$.

Results

Patients

Seventeen patients were included in this study (Table 1). They were affected by hemangioma in 10 cases (59%), schwannoma in 6 cases (35%), and meningioma in 1 case (6%). The mean age was 43 ± 12.9 years (range = 22-68). The tumors were located in the right temporal bone in 8 cases (47%) and in the left in 9 cases (53%). There was no

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<tr>
<th>Patient</th>
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<th>POST</th>
<th>PRE</th>
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<td>III</td>
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<td>IV</td>
<td>III</td>
<td>10</td>
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<td>2.6</td>
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</table>

Abbreviations: FNG, facial nerve graft; HB, House and Brackman; Hem, hemangioma; LFU, lost to follow-up; Men, meningioma; POST, postoperative; PRE, preoperative; PTA, pure-tone audiometry; Schw, schwannoma.
difference between the groups in side of the tumor, sex, or age (Fisher and Wilcoxon tests).

**Preoperative Data**

All patients presented some degree of facial impairment as first symptoms. The overall facial function was grade II in 1 case (6%), grade III in 1 case (6%), grade IV in 8 cases (47%), grade V in 4 cases (23.5%), and grade VI in 3 cases (17.5%). Preoperative FN function was not related to tumor size, patient age, or preoperative duration of facial palsy (Kruskal-Wallis test). However, severe FN palsy (grade V or VI) was observed only in hemangioma cases: 7 patients with hemangioma (70%) had HB V or VI and no patients with schwannoma ($P = .01$, Fisher test; Table 2). The pattern of facial palsy was progressive in 11 cases (65%), sudden for 4 (23.5%), and recurrent in 2 (12%). There was no significant difference among these modes of evolution depending on the type of tumor (Fisher test, data not shown).

Regarding the hearing status, an overall hearing impairment was present in 4 cases (23.5%), 3 schwannomas and 1 hemangioma, which were 3 cases of sensorineural hearing loss and 1 case of conductive hearing loss. PTA was significantly higher in the case of schwannoma versus hemangioma ($P = .007$, Wilcoxon test; Table 2), but a significant correlation between tumor size and PTA was found (Pearson test, $r = 0.8$, $P < .0001$). There was no difference in vestibular impairment per tumor type (Table 2) or tumor size (Fisher test).

Regarding the size of the tumor (Table 2), hemangiomas were significantly smaller versus schwannomas: the mean diameters were $8 \pm 3.7$ mm and $16 \pm 7.3$ mm for hemangioma and schwannoma, respectively ($P = .01$, Wilcoxon test). Two patients had a preoperative cochlear fistula identified on preoperative imaging studies (cases 12 and 16; Table 1).

**Surgery**

Regarding the surgical approach, the middle cranial fossa was used in all cases. Computer-assisted surgical navigation and FN-stimulating burr were used since 2007 and 2010, respectively. A total tumor resection was achieved in all cases; the FN was interrupted and immediately repaired in 16 patients (94%), with a great auricular nerve graft in 15 cases and a sural nerve graft in 1 case (due to the small diameter of the great auricular nerve). One patient (6%) underwent a total resection of a GG schwannoma with an anatomically intact FN at the end of the procedure. In this particular case, a good dissection plane could easily be found, and the tumor could be separated for the FN (case 14; Table 1).

**Postoperative Outcomes**

There were no major complications: no temporal lobe injuries, no cerebrospinal fluid leaks. One case of asymptomatic postoperative extradural hematoma with spontaneous resolution was observed.

The mean follow-up was $3.7 \pm 2.99$ years (range = 1-11; $n = 16$). One patient was lost to follow-up 1 month after surgery. At the last postoperative consultation, the overall facial function ($n = 16$) was grade III in 11 cases (69%), grade IV in 4 cases (25%), and grade V in 1 case (6%). Facial function improved in 12 cases (75%), stabilized in 3 (19%), and worsened in 1 (6%), from HB II to HB III. Regarding hemangioma and schwannoma cases that had FN interruption with an FN graft ($n = 14$), the overall facial function at last postoperative consultation was grade III in 9 cases (64%), grade IV in 4 cases (29%), and grade V in 1 case (7%). Facial function improved in 10 cases (71%), stabilized in 3 cases (21%), and worsened in 1 case (7%), from HB II to HB III. Severe synkinesis were reported for 2 patients (14%) who had HB IV. There was no difference in FN outcomes when hemangioma cases were compared with schwannoma cases (Table 1; Fisher test).

Age, tumor size, and duration of preoperative facial palsy were not related to FN function at 12 months postoperatively (Kruskal-Wallis test). Conversely, preoperative HB VI was significantly related to poorer postoperative FN function after FN grafting in patients who had surgery for Geniculate Ganglion Tumor.

### Table 2. Preoperative Clinical Features of 17 Patients Who Underwent Surgery for Geniculate Ganglion Tumor.

<table>
<thead>
<tr>
<th></th>
<th>Total (N = 17)</th>
<th>Hemangioma (n = 10)</th>
<th>Schwannoma (n = 6)</th>
<th>Meningioma (n = 1)</th>
<th>P Value</th>
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</thead>
<tbody>
<tr>
<td>Size, mm</td>
<td>$11 \pm 6.3$</td>
<td>$8 \pm 3.7$</td>
<td>$16 \pm 7.3$</td>
<td>$5$</td>
<td>.03$^b$</td>
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<tr>
<td>FN function</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Grade II</td>
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<td>1</td>
<td>0</td>
<td>.4</td>
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<tr>
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<td>0</td>
<td>0</td>
<td>1</td>
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<tr>
<td>Grade IV</td>
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<td>2</td>
<td>5</td>
<td>1</td>
<td>.03$^b$</td>
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<tr>
<td>Grade V</td>
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<td>4</td>
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<td>0</td>
<td>.2</td>
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<tr>
<td>Grade VI</td>
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<td>3</td>
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<td>0</td>
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</tr>
<tr>
<td>PTA, dB</td>
<td>$21 \pm 28$</td>
<td>$10 \pm 8.7$</td>
<td>$41 \pm 41$</td>
<td>$10$</td>
<td>.007$^b$</td>
</tr>
<tr>
<td>VI</td>
<td>7</td>
<td>2</td>
<td>4</td>
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<td>.1</td>
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<td>Tinnitus</td>
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</table>

Abbreviations: FN, facial nerve; PTA, pure-tone audiometry; VI, vestibular impairment.

$^a$Values presented as n or mean $\pm$ SD.

$^bP < .05$ (Fisher and Wilcoxon tests, hemangioma vs schwannoma).
hemangioma or schwannoma (n = 14); indeed, a worse postoperative FN function (grade IV, V, VI) was present in all patients who had preoperative HB VI (n = 3) and in only 18% (n = 2) of patients who had less severe preoperative FN function (n = 11; \( P = .02 \), Fisher test).

Regarding the hearing, it was preserved in 15 patients (94%) and worsened in 1 (6%; from class C to class D), who was affected by a cochlear fistula. Postoperative MRI was available for 16 patients, and no recurrence was detected in yearly performed MRI.

**Discussion**

This study demonstrates that although the main symptom is facial palsy for all types of GG tumors, facial impairment is more severe in cases of hemangioma than in cases of schwannoma. Severe FN palsy in cases of hemangioma has been reported in other studies, but this study is the first to compare these 2 tumor types.

Several hypotheses were highlighted in the literature to explain FN palsy in GG tumors: the compression by the growing tumor is certainly one of the main factors, followed by invasion of the nerve as identified by histologic analysis. Moreover, since hemangioma is a vascular malformation that develops from the rich venous plexus that surrounds the GG, some authors raised the hypothesis of vascular steal, which causes facial palsy by an ischemic phenomenon for this type of tumor. This could account for the more severe FN impairment for hemangioma that is usually smaller at diagnosis when compared with schwannoma, as reported in other studies. These clinical differences, with radiologic features (Table 3, Figure 1), could help in making the correct diagnosis.

The present study describes only cases of total excision. It shows that postoperative outcomes did not depend on tumor histology for GG tumors. When surgery is indicated, postoperative FN function improves in most patients, with the majority reaching postoperative HB III (64% of cases), which is in line with results reported in the literature: HB III for 55% to 86% after FN grafting. Synkinesis is very difficult to assess, but the low incidence of severe synkinesis in the present study could be explained by the use of short grafts—from the labyrinthine to tympanic portions of the FN (tumors located only on the GG)—which could contribute to more precise axonal regrowth. Complete resection remains the curative treatment for GG tumor, and its indication depends on preoperative FN function. Indeed, since FN interruption is necessary in most cases to achieve complete resection, most authors advocate surgery when FN function is at least HB III or worse. A conservative approach with dissection of the FN was possible in 1 schwannoma, as proposed by others. Such a result was not achieved for hemangiomas due to tumor invasion of the

| Table 3. Radiologic Features for the 3 More Frequent GG Tumors |
|------------------|------------------|------------------|
|                   | Schwannoma       | Hemangioma       | Meningioma       |
| CT scan           | Smooth walled, homogeneous hypodensity round or oval shaped, hourglass aspect | Irregular margins, intratumoral calcifications with honeycomb appearance | Irregular margins, rare intratumoral calcification |
| CT scan with contrast MRI | Irregular enhancement | Avid enhancement | Avid enhancement |
| T1                | Iso- or hypointense | Isointense | Iso- or hypointense |
| T2                | Iso- or hyperintense | Hyperintense | Iso- or hyperintense |
| T1 with contrast   | Irregular avid enhancement | Avid enhancement | Avid enhancement |

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.

Figure 1. T1 with contrast magnetic resonance image showing geniculate ganglion tumors: (A) hemangioma, (B) meningioma, and (C) schwannoma.
FN, although some cases have been reported in literature. In case of meningioma, only 1 report with dissection of the FN from the tumor has been published.

Regarding hearing, surgery allows preservation in most cases through a middle cranial fossa approach that is routinely used for GG tumors. As showed in this study, preoperative cochlear fistula could be associated with a worsening of hearing.

Other options can be proposed for the management of GG tumors. The first is the wait-and-scan strategy, which can be a good option in the case of a nongrowing poor symptomatic tumor, with normal or near-normal FN function (HB I and II). These tumors are more frequently schwannomas than hemangiomas because of more severe facial function in cases of the latter, as seen previously. In a review of 120 cases of GG hemangiomas, only 11 had been observed, and the facial function remained stable for only 28% of observed cases. The second option is decompression surgery that aims to avoid the axonal lesions of the FN caused by tumor compression in the fallopian canal. Wilkinson et al reported an improvement of FN function in 16% of cases and a decrease in 21% of cases for 21 patients who had decompression surgery for FN schwannoma, with no difference in tumor evolution between decompression and observation. Decompression can be a good option when the tumor is confined in the fallopian canal, but most of the time, when the tumor is located only on the GG, the bony roof has already been eroded by the tumor itself.

Radiosurgery is a viable option in case of growing schwannoma of the GG with FN function grade I or II. The goal of radiosurgery is to reduce or stabilize the tumor volume and the FN function. According to the literature, the tumor size is stabilized or reduced in 83% to 100% of patients, and FN function is improved or stabilized in 67% to 100%. Regarding patients’ hearing after radiosurgery, a meta-analysis of 14 patients treated with radiosurgery for FN schwannoma for whom the auditory data were available reported worsened hearing for 36.7% of patients. Concerning hemangiomas, no studies have been published on the use of radiosurgery for these tumors. So, in case of preoperative GG tumors with a good preoperative FN function (grade I or II), to avoid unnecessary and ineffective treatment, establishing a correct diagnosis of the tumor type is fundamental because only schwannoma could be successfully treated with radiosurgery. In the other cases, a wait-and-scan policy is a viable option.

Limitations of the study include its retrospective nature and the poor statistical power due to the small sample size. This is a result of the scarcity of GG tumors. This also precludes the ability to perform multivariate analysis for prognosis factors assessment. Finally, the assessment of FN function can be discussed as a limitation because of the subjectivity of the HB scale. This scale is not good for synkinesis and spasm evaluation, as a patient can be assessed as HB III or IV based solely on the severity of his or her spasms and synkinesis. Nevertheless, it is the more common scale used by neurotologists in the literature, and results of this study are comparable to other postoperative outcomes in terms of FN function.

**Conclusion**

Hemangioma appears to be smaller but more aggressive than schwannoma on FN function. Establishing the correct diagnosis is mandatory for choosing the appropriate management, and when surgery is indicated, this option is safe and effective with few complications and no recurrences.

**Author Contributions**

Ghizlene Lahhou, conception and design of the work, acquisition, analysis and interpretation of the data, drafting the work, final approval of the version to be published, agreement to be accountable for all aspects of the work; Yann Nguyen, contribution to design of the work, substantial contribution to acquisition of the data, revising the work, final approval of the version to be published, agreement to be accountable for all aspects of the work; Francesca Yoshiie Russo, contribution to design of the work, substantial contribution to analysis of the data, revising the work, final approval of the version to be published.
approval of the version to be published, agreement to be accountable for all aspects of the work; Evelyne Ferrary, contribution to design of the work, substantial contribution to statistical analysis of the data, revising the work, final approval of the version to be published, agreement to be accountable for all aspects of the work; Olivier Sterkers, conception and design of the work, contribution to analysis and interpretation of the data, revising the work, final approval of the version to be published, agreement to be accountable for all aspects of the work; Daniele Bernardeschi, conception and design of the work, interpretation of data for the work, revising the work, final approval of the version to be published, agreement to be accountable for all aspects of the work.

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