Geniculate Ganglion Tumors: Clinical Presentation and Surgical Results

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Keywords: Geniculate ganglion; hemangioma; schwannoma; meningioma; facial nerve graft

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 of 17 patients operated on for a geniculate ganglion tumor removal (10 hemangiomas, 6 schwannomas, one meningioma) were reviewed. 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 grade V and VI) was present in 70% of cases for hemangiomas and for no case of schwannoma ($p = 0.01$), although hemangiomas were significantly smaller tumors ($p=0.01$). Hearing loss was observed in 4 cases (23.5%), and was related to tumor volume ($p<0.0001$). 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-Brackman 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. Sugery, when indicated, is safe and effective, and postoperative outcomes are not related to tumor type.

INTRODUCTION

Facial nerve (FN) tumors are rare lesions of the petrous bone.[1] They include schwannomas, hemangiomas, and meningiomas. Schwannomas represent 0,8% of petrous bone tumors.[2] It is a benign, encapsulated, slow growing lesion arising from the Schwann cells, that can involve any of the different segments of the FN, the geniculate ganglion (GG) being the most frequent segment involved.[3] FN hemangiomas encompass 0.7% of petrous bone tumors [4] and they have been nowadays reclassified in the group of vascular malformations.[5] As schwannomas, hemangiomas are mainly located in the GG.[5] FN meningiomas are an extremely rare tumor, with only some case reports reported in the literature.[6,7] They arise from arachnoid cells accompanying the FN during its embryonic formation.[8] The GG and the internal auditory canal are the two preferential locations of petrous bone meningiomas.[7]

When the tumor is limited to the GG, the most common symptom is facial paralysis. Even if the radiological features of these lesions (MRI and CT-scan) might help in the differential diagnosis, no study has been done to analyze differences in clinical presentation among the different tumor types that could help in establishing a correct preoperative diagnosis. This is nowadays essential in order to define the correct management of these tumors considering the increasing number of patients affected by FN schwannoma treated with stereotaxic radiosurgery.[9]

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

Methods

Medical records of patients operated on for an intrapetrous FN tumor from 1988 to 2013 period in a tertiary referral center were retrospectively reviewed. All patients gave their informed consent 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 (OS), for preoperative 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 post-operative histological examination. Tumors located in another segment of the FN or spreading from the GG to another segment were excluded. Non-primary 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 histological examination after tumor excision. Frozen sections of the facial nerve at proximal (labyrinthine) and distal (tympanic) segment of the FN were performed in all but one patient who underwent tumor excision without interruption of the FN.

Pre-operative Assessment

The data at the first consultation included demographic information, presenting symptoms, facial function according to the House and Brackman (HB) scale,[10] the mean pure-tone audiometry (PTA) (mean of 500,1000, 2000 and 3000 Hz) in air and bone conduction with headphones, hearing classification according to AAO-HNS, and vestibular function evaluated by a caloric test. A vestibular impairment was defined as a lateralization and a directional preponderance more than 25% calculated by Jongkee's formula. Imaging (CT scan and magnetic resonance imaging – MRI - in T1-WI, T2-WI, and T1 with contrast) was available

for all patients and was analysed for tumor size on the post–contrast T1-WI sequences, and the presence of cochlear or labyrinthine fistula on CT-scan and T2-WI.

Peri- and Post-operative 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 in the three first postoperative years of follow-up (FU).

Statistical Analysis

Results are presented as mean ± standard deviation (SD). Hemangiomas and schwannomas were compared using Ficher tests for qualitative data; Wilcoxon tests, Kruskall-Wallis tests and Pearson tests for quantitative data. Statistical tests were performed using R (version 3.2.3). Differences were considered statistically significant when $p < 0.05$.

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 one case (6%). The mean age was 43 ± 12.9 years (range = 22 - 68). The tumours were located in the right temporal bone in 8 cases (47%) and in the left in 9 cases (53%). There was no 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 one case (6%), grade III in one case (6%), grade IV in 8 cases (47%), grade V in 4 cases (23.5%), and grade VI in three cases (17.5%). Preoperative FN function was not related to tumor size, to patient age, or to preoperative duration of facial palsy (Kruskall-Wallis test). However, severe FN palsy (grade V or VI) was observed only in hemangioma cases: 7 patients with hemangioma (70%) had a HB V or VI and no patients with schwannoma ($p=0.01$, Fisher test – **Table 2**)

The pattern of the facial palsy was progressive in 11 cases (65%), sudden for 4 patients (23.5%), and recurrent in two cases (12%). There was no significant difference between these different 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%), which were three schwannomas and one hemangioma, which were three cases of sensorineural hearing losses and one case of conductive hearing loss. The PTA was significantly higher in case of schwannoma compared to hemangioma ($p=0.007$ – Wilcoxon test, **Table 2**) but a significant correlation between tumor size and PTA was found (Pearson test, $r=0.8$, $p<0.0001$).

There was no difference in vestibular impairment depending on tumor type (**Table 2**) or tumor size (Fisher test).

Regarding the size of the tumor (**Table 2**), hemangiomas were significantly smaller compared to schwannomas: the mean diameter was 8 ± 3.7 mm and 16 ± 7.3 mm for hemangioma and schwannoma respectively ($p=0.01$ – Wilcoxon test). Two patients had a pre-operative cochlear fistula identified on pre-operative imaging studies (case 12 and 16, **table 1**).

Surgery

Regarding the surgical approach, the middle cranial fossa was used in all cases. Computer-assisted surgical navigation [11] and the facial nerve stimulating burr [12] 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%), using a great auricular nerve graft in 15 cases and a sural nerve graft (due to the small diameter of the great auricular nerve) in one case. 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 be found easily 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 cerebro-spinal fluid leaks. One case of asymptomatic postoperative extradural hematoma with spontaneous resolution was observed.

The mean follow-up was 3.7 ± 2.99 years, range = 1 – 11 (n=16). One patient was lost to follow-up one 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 one case (6%). Facial function improved in 12 cases (75%), stabilized in 3 cases (19 %) and worsened in one case (6%) from a HB II to a HB III. Regarding hemangiomas and schwannomas' cases that had FN interruption with a 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 one case (7%). Facial function improved in 10 cases (71%), stabilized in 3 cases (21 %) and worsened in one case (7%) from a HB II to a HB III. Severe synkinesis were reported for two patients (14%) who had a HB IV. There was no difference in FN outcomes comparing hemangioma cases to schwannoma cases (**Table 1**, Fisher test).

Patient's age, tumor size and duration of preoperative facial palsy were not related to FN function at 12 months postoperatively (Kruskall-Wallis test). Conversely, preoperative HB grade VI was significantly related to poorer post-operative FN function after FN grafting in patients operated for hemangioma or schwannoma (n=14): indeed, a worse postoperative FN

function (grade IV, V, VI) was present in all patients who had a preoperative HB VI (n=3), and only in 18% (n=2) of patients who had less severe preoperative FN function (n=11) (p=0.02 – Fisher test).

Regarding the hearing, it was preserved in 15 cases (94%) and worsened in one patient affected by a cochlear fistula from a class C to a class D (6%).

Post-operative MRI was available for 16 patients and no recurrence was detected in a 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 already been reported in other studies, [13–15] but this is the first study that compares theses two tumor types.

Several hypotheses were highlighted in literature to explain the FN palsy in GG tumors: the compression by the growing tumor [7,16] is certainly one of the main factors followed by the invasion of the nerve identified by histological analysis.[14] Moreover, since hemangioma is a vascular malformation that develops from the rich venous plexus that surround the GG,[17] some authors raised the hypothesis of a vascular steal that causes a facial palsy by an ischemic phenomenon for this type of tumor.[18] This could account for the more severe FN impairment for hemangioma that is usually smaller at diagnosis compared to schwannoma, as already reported in other studies.[13,19] These clinical differences, together with radiological 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, the postoperative FN function improves in most of the patients with the majority of patients

reaching a postoperative HB grade III (64% of cases), which is in line with results reported on literature: from 55% to 86% of HB III after facial nerve grafiting.[20–22] Synkinesis are very difficult to asses, but the low incidence of severe synkinesis in the present study could be explained by the use of short grafts, from labyrinthine to tympanic portion of the facial nerve (tumors located only on the geniculate ganglion), that could contribute to a more precise axonal regrowth. Only a preoperative HB grade VI doesn't provide a satisfactory recovery of the nerve. This has been already been pointed out in another studies.[20,22] The complete tumor resection remains the curative treatment for GG tumors, and its indication depends on the preoperative facial nerve function.[3] Indeed, since a FN interruption is necessary in most of the cases to achieve a complete resection, most of the authors advocate surgery when the FN function is at least HB III,[3,20,21,23] or worse. A conservative approach with dissection of the FN was possible in one schwannoma as proposed by others.[24,25] Such a result was not achieved for hemangiomas due to the tumor invasion of the FN,[13] although some cases have been reported in literature.[13,26] In case of meningioma, only one report with a dissection of the FN from the tumor has been published.[27]

Regarding hearing, surgery allows hearing preservation in most of the cases through a middle cranial fossa approach, that is routinely used for GG tumors.[15,17,19] As showed in this study, preoperative cochlear fistula could be associated to a worsening of hearing.[13,14]

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 case of a non-growing poor symptomatic tumor, with a normal or near-normal FN function (HB I and II). These tumors are more frequently schwannomas than hemangiomas because of a more severe facial function in cases of hemangiomas as seen previously. In a review of 120 cases of GG hemangiomas, only 11 had been observed, and the facial function remained stable only for 28% of observed cases.[15] The second option is a decompression surgery that aims to avoid the axonal lesions of the FN caused by the tumor compression in the Fallopian canal. Wilkinson et al reported an

improvement of the FN function in 16% of cases and a decrease in 21% of cases for 21 patients who had a decompression surgery for a FN schwannoma, with no difference in tumor's evolution between decompression and observation.[20] 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 a FN function grade I or II. The goal of radiosurgery is to reduce or to stabilize the tumor volume and the facial nerve 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% of patients.[20,28–31] 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 36.7% of patients whose hearing worsened.[9] Concerning hemangiomas, no studies have been yet 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), in order 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, 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 the result of GG tumor's scarcity. This also enables the ability to perform multivariate analysis for prognosis factors assessment. Finally, the assessment of FN function can be discussed because of the subjectivity of HB scale. Also, this scale is not a good scale for synkinesis and spasm evaluation, as a patient can be assessed on a HB III or IV regarding the severity of his spasms and synkinesis. Nevertheless, it is the more common scale used by neurotologists in literature, and results of this study are comparable to other postoperative outcomes in terms of FN function.[20,21]

Conclusion

Hemangioma appears to be smaller in size but more aggressive on FN function than schwannoma. Establishing the correct diagnosis is mandatory for choosing the appropriate management (**Figure 2**) and, when surgery is indicated, this option is safe and effective with few complications and no recurrences.

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Patient	Age (year)	Tumor	Tumor size (mm)	Duration of FN palsy (months)	FN management	HB pre	HB post	PTA pre (dB)	PTA post (dB)	FU (year)
1	46	Hem	7.7	4	FNG	IV	III	11	16	1.5
2	28	Hem	4	12	FNG	V	III	10	7	3.3
3	46	Hem	8	15	FNG	III	III	10	14	1.3
4	22	Hem	15	22	FNG	V	III	5	9	3.4
5	58	Hem	13	24	FNG	VI	IV	10	10	3.9
6	53	Hem	6	32	FNG	V	*LFU*	5	*LFU*	*LFU*
7	32	Hem	5	39	FNG	VI	IV	5	10	1.2
8	30	Hem	10	50	FNG	V	III	34	41	11.2
9	47	Hem	4	60	FNG	VI	V	5	15	1.3
10	55	Hem	9	250	FNG	IV	III	5	10	2
11	35	Schw	15	2	FNG	II	III	20	20	4.9
12	68	Schw	13	5	FNG	IV	IV	53	101	1
13	24	Schw	10	8	FNG	IV	III	15	18	5.4
14	41	Schw	15	12	Dissection	IV	III	24	21	9.3
15	50	Schw	11	26	FNG	IV	III	16	16	2
16	37	Schw	30	179	FNG	IV	IV	120	120	5.9
17	48	Men	5	44	FNG	IV	III	10	10	2.6

Table 1: Patients' characteristics.

HB = House and Brackman; Schw = schwannoma; Hem = hemangioma; Men = meningioma; FNG = Facial nerve graft; FU = follow up; LFU = lost to follow-up

		Total *N=17*	**Hemangioma N=10**	**Schwannoma N=6**	**Meningioma N=1**	**p**
Size (mm) (mean ± SD)		*11 ± 6.3*	8 ± 3.7	16 ± 7.3	5	0.03*
FN function	Grade II	*1*	0	1	0	0.4
	Grade III	*1*	1	0	0	1
	Grade IV	*8*	2	5	1	0.03*
	Grade V	*4*	4	0	0	0.2
	Grade VI	*3*	3	0	0	0.2
PTA (dB) (mean ± SD)		*21 ± 28*	10 ± 8.7	41 ± 41	10	0.007*
Vestibular impairment		*7*	2	4	1	0.1
Tinnitus		*2*	0	2	0	0.2

Table 2: Preoperative clinical features of 17 patients who underwent surgery for GG tumor.

PTA = Pure-tone audiometry; * = significant (Fisher and Wilcoxon tests comparing hemangioma to schwannoma).

		Schwannoma	Hemangioma	Meningioma
CT scan		Smooth-walled Homogenous hypodensity round or oval-shaped Hourglass aspect	Irregular margins Intratumoral calcifications with honeycomb appearance	Irregular margins Rare intratumoral calcification
CT scan with contrast		Irregular enhancement	Avid enhancement	Avid enhancement
MRI	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

Table 3: Radiological features for the three more frequent GG tumors [6,32,33]

Figures Legends

Figure 1: T1 with contrast MRI showing a GG hemangioma (A), a GG meningioma (B), and a GG schwannoma (C).

Figure 2: Management of geniculate ganglion tumors depending on the tumor type. HB = House and Brackman.

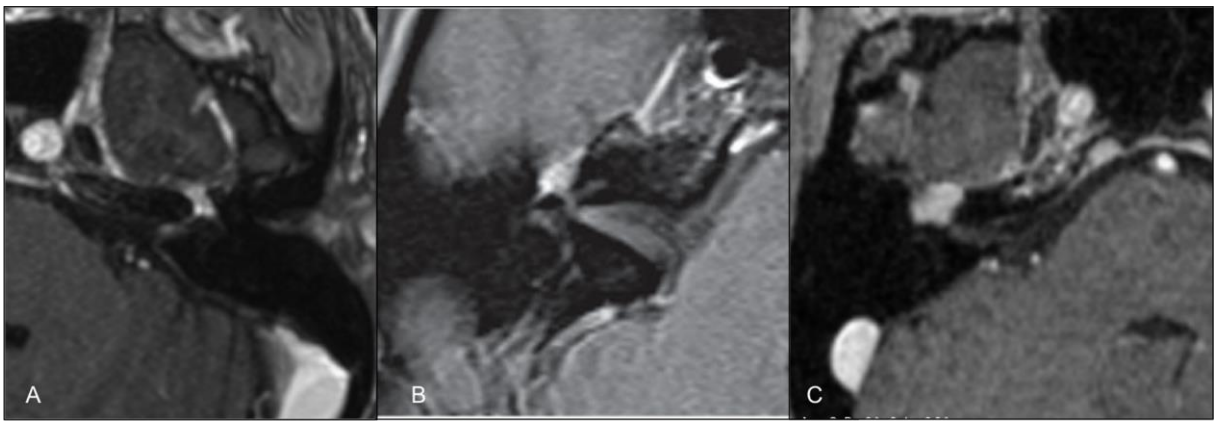

Fig. 1

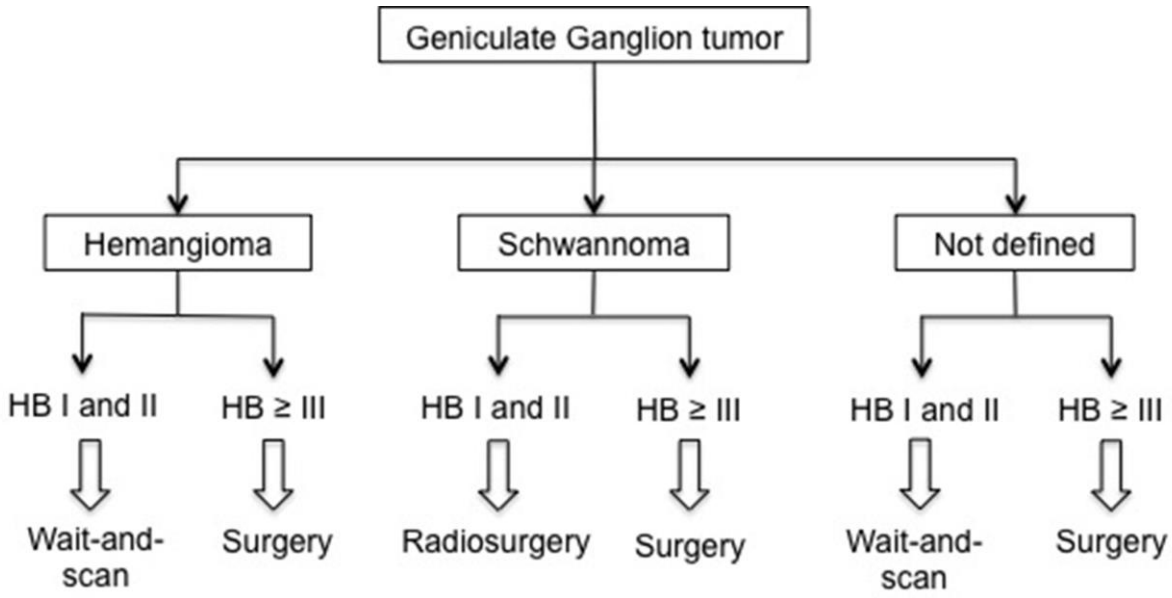
Geniculate Ganglion tumor
Hemangioma
Schwannoma
Not defined
HB I and II
HB ≥ III
HB I and II
HB ≥ III
HB I and II
HB ≥ III
Wait-and-scan
Surgery
Radiosurgery
Surgery
Wait-and-scan
Surgery

Fig. 2