European Archives of Oto-Rhino-Laryngology
https://doi.org/10.1007/s00405-021-06765-x

OTOLOGY

Management of facial nerve schwannoma: when is the timing for surgery

Young Sang Cho[1] · Ji Eun Choi[2] · Ji Hyun Lim[3] · Yang-Sun Cho[1]

Received: 18 September 2020 / Accepted: 15 March 2021

Abstract

Purpose Although the estimated prevalence is extremely low, facial nerve schwannoma (FNS) is the most common primary tumor of the facial nerve (FN). In the present study, the outcome of surgical management in 18 patients with FNS was analyzed and an appropriate time for surgery was proposed.

Materials and methods A total of 18 patients with FNS who underwent surgical management by a single surgeon from 1999 to 2018 were retrospectively analyzed.

Results Among the 18 patients, five had no facial paralysis before surgery. Near-total removal was performed in three cases, and two cases were managed with decompression. In 13 cases with various degree of preoperative facial palsy, nerve continuity was lost during surgery. FN was reconstructed using cable graft in ten cases, direct anastomosis in one case, and facial-hypoglossal nerve transfer in one case. Facial reanimation surgery without FN reconstruction was performed in one case due to a long-standing facial paralysis before surgery. Preoperative House-Brackmann (H-B) grade in all patients was significantly worse as tumor size increased. The correlation was not observed between the duration and severity of preoperative facial palsy.

Analysis of 12 patients who underwent FN reconstruction revealed that all patients with good preoperative facial function (H-B grade II–III) recovered to H-B grade III after surgery (7/7, 100%). However, patients with poor preoperative facial function (H-B grade IV or worse) had only a 40% (2/5) chance of improving to grade III after surgery. Preoperative tumor size and duration of facial palsy did not affect postoperative final facial function.

Conclusion We suggest that H-B grade III facial palsy is the best time for surgical intervention, regardless of the tumor size or duration of facial palsy.

Keywords Facial nerve schwannoma · Facial palsy · House-Brackmann grade · Facial nerve function · Surgical treatment

Introduction

Facial nerve schwannoma (FNS) is the most common primary tumor of the facial nerve (FN), although the estimated prevalence is extremely low [1]. Similar to vestibular schwannoma (VS), FNS is a benign tumor originating from Schwann cells that are slow-growing and may be present for years before symptoms arise [2]. The signs and symptoms of FNS depend on the anatomical origin and extent. The most common clinical manifestation is progressive FN dysfunction due to compression by the mass, often combined with hearing loss, pain, vertigo, and less frequently, a parotid mass [3]. The total number of cases reported in the literature is just over 600 cases to date. Because the majority of published case studies include a relatively small number of patients and variable treatment outcomes, clear guidelines for the management of FNS have not been established [4].

The management of FNS is particularly challenging and several options exist. 'Wait and see policy' is a viable option because the tumor is extremely slow-growing and often diagnosed in patients with normal facial function [3].

✉ Yang-Sun Cho
yscho@skku.edu

[1] Department of Otorhinolaryngology-Head and Neck Surgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, 81 Irwon-ro, Gangnam-gu, Seoul 06351, South Korea

[2] Department of Otorhinolaryngology-Head and Neck Surgery, Dankook University Hospital, Cheonan, South Korea

[3] Center for Clinical Epidemiology, Samsung Seoul Hospital, Seoul, South Korea

Published online: 08 April 2021

Management options include surgery and radiation therapy. Complete surgical resection is the only cure; however, the results of surgical resections vary widely [5–7]. The fascicle preservation surgery shows better functional outcome but is possible in only a limited number of cases [8]. Although the effects of radiation therapy have been reported in recent years, the results of stereotactic radiosurgery (SRS) show inconsistent outcomes with regard to FN and hearing outcomes, mainly because some patients underwent conservative surgical management including decompression before or after SRS [9, 10].

The timing of the surgery is the most critical issue in the surgical management of FNS. If the patient has severe to total facial paralysis, tumor resection and FN reconstruction should be attempted as soon as possible because the final outcome is dependent on the duration of facial palsy [11–15]. However, many patients visit the clinic only after the development of obvious facial palsy, and some patients often present with other neurotologic symptoms and normal to near normal facial function [16].

Regarding the timing of surgery, the consensus is to perform surgery at House-Brackmann (H-B) grade III [17] or grade IV [18]. However, details of tumor extent were lacking in those studies and many patients with preoperative normal facial function were included.

In the present study, the outcomes of 18 patients with a variable preoperative facial function who underwent surgical intervention by a single surgeon, and who were followed up for more than one year were analyzed. In addition, the most appropriate time for surgical management was discussed.

Materials and methods

Subjects

The medical records of patients who were treated for FNS from 1999 to 2018 in a tertiary referral center were retrospectively reviewed. FN tumors other than schwannoma (e.g., glomus faciale, neurofibroma) were excluded. Overall, 18 patients (7 males and 11 females) who were followed up for more than 14 months were included in the study. The mean age was 39.94 years (range 23–67 years) and the mean follow-up period was 43.3 months (range 14.7–68.8 months). This study was approved by the Institutional Review Board of the Samsung Medical Center and followed the declaration of Helsinki guidelines (IRB File No. 2019-03-065).

A single neurotologist evaluated and operated on all patients and patient demographics are presented in Table 1. Preoperatively, temporal bone computed tomography (TBCT) and internal auditory canal magnetic resonance imaging (IAC-MRI) scans were taken. The tumor size was measured based on the longest axis on the MRI. The facial function was evaluated using the H-B grading scale [19]. After surgery, H-B grade and IAC-MRI were assessed at regular follow-ups.

Tumor removal and FN reconstruction

Various surgical approaches were used based on the location and size of the tumor, residual hearing, and vestibular functions (Table 1). During the operation, FN monitoring was conducted continuously using the electrophysiological response (NIM 3.0 monitor, Medtronic, Jacksonville, FL, USA).

Nerve fascicle preservation surgery was attempted in some cases; however, removal of the tumor while preserving the FN continuity was not possible. Nerve reconstruction was performed immediately when nerve continuity was lost during the operation. All reconstructions were performed with three epineurium sutures (nylon 9-0) using microscopic techniques. Direct end-to-end anastomosis was performed in one case (case #2) where the proximal portion of FN at the brainstem was available, and the cut ends could be anastomosed after rerouting the tympanic and mastoid portion of the FN. If direct anastomosis was not possible, reconstruction was performed with cable graft using a sural nerve or great auricular nerve (GAN). If the duration of complete paralysis was long enough, hypoglossal-facial nerve transfer or facial reanimation surgery was performed.

Statistical analysis

SAS version 9.4 (SAS Institute Inc., Cary, NC, USA) was used for all statistical analyses and a p value < 0.05 was considered statistically significant. The Fisher's exact test was used to analyze the effect of preoperative H-B grade on postoperative outcome. The Spearman analysis was used to determine whether preoperative H-B grade correlates with tumor size or symptom duration.

Results

Preoperative facial function status

The preoperative H-B grade was relatively good in 12 patients (I–III) and 6 patients had a poor facial function grade IV–VI. The duration of facial palsy ranged from 2 to 300 months. The mean follow-up duration after surgery was 43.3 months and the H-B grade evaluated at the last follow-up was used for analysis.

Analysis of preoperative H-B grade, the tumor size, and duration of facial palsy showed the preoperative H-B grade was significantly worse as tumor size increased

Table 1 Demographic characteristics and surgical methods

No	Sex/ Age	Symptoms (FNP duration, months)	Tumor size, mm	Tumor removal	Surgical approach	FN management	H-B grade (preoperative/1 month/6 months/last)	Follow-up duration (months)
1	F/62	FNP (7)	18	GTR	Transmastoid	Cable graft with GAN	3/6/6/3	64.8
2	F/27	FNP (4)	21	NTR	Translabyrinthine	Direct anastomosis	2/4/4/3	68.8
3	F/36	FNP (60)	17	GTR	Transmastoid	CN XII-VII, hook-up	5/4/4/4	14.7
4	F/50	HL	7	NTR	MFA	N/A	1/6/3/3	74.1
5	F/51	HL, FNP (4)	4	GTR	Combined approach (MFA + transmastoid)	Cable graft with sural nerve	4/6/3/3	42.1
6	F/49	Vertigo	7	NTR	Translabyrinthine	N/A	1/6/3/3	73.0
7	M/34	FNP (2)	23	GTR	Transmastoid	Cable graft with sural nerve	2/4/4/3	58.3
8	M/23	HL	12	NTR	Translabyrinthine	N/A	1/4/3/3	29.5
9	M/28	HL	9	DEC	Transmastoid	N/A	1/1/1/1	29.2
10	F/67	HL, FNP (228)	35	GTR	Transmastoid	Facial reanimation	6/6/6/6	23.1
11	F/35	HL	10	DEC	Translabyrinthine	N/A	1/1/1/1	59.5
12	M/40	HL, FNP (120)	28	GTR	Transmastoid	Cable graft with sural nerve	4/6/6/4	17.8
13	F/33	HL, FNP (27)	4	GTR	Combined approach (MFA + transmastoid)	Cable graft with sural nerve	3/6/6/3	49.3
14	M/30	FNP (36)	28	GTR	Transmastoid	Cable graft with sural nerve	6/6/5/5	55.5
15	M/15	FNP (18)	25	GTR	Transmastoid	Cable graft with sural nerve	3/4/4/3	48.8
16	M/54	FNP (120)	16	GTR	Translabyrinthine	Cable graft with sural nerve	3/5/5/3	41.6
17	F/41	FNP, HL (300)	19	GTR	Combined approach (MFA + transmastoid)	Cable graft with sural nerve	3/6/4/3	24.0
18	F/55	FNP, HL (28)	18	GTR	Combined approach (MFA + transmastoid)	Cable graft with sural nerve	4/6/4/3	17.4

FNP facial nerve palsy, *HL* hearing loss, *GTR* gross total resection, *NTR* near total resection, *DEC* decompression, *MFA* middle fossa approach, *CN* cranial nerve, *GAN* great auricular nerve

($p = 0.029$). However, the correlation between the duration of preoperative facial palsy and H-B grade did not show statistical significance (Fig. 1).

The tumor size ranged from 4 to 35 mm. The tumor extents were very diverse, including the cases confined within the IAC as well as cases extending to the parotid gland (Table 2).

Surgical methods and tumor extent

Five patients (27.8%) had a normal preoperative facial function; four had a tumor confined to the IAC, which was diagnosed as VS. Three patients (Table 1, patients #4, #6, #8) underwent near-total removal of the tumor and developed facial paralysis H-B grade IV–VI immediately after surgery

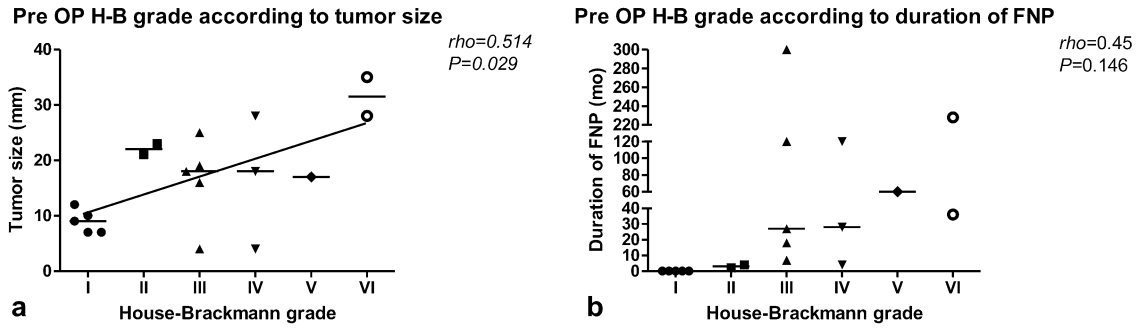

Fig. 1 Preoperative House-Brackmann (H-B) grade based on preoperative tumor size and duration of facial nerve paralysis (FNP). The larger the tumor size, the worse the H-B grade before surgery (**a**). The duration of facial palsy did not reach statistical significance (**b**)

Table 2 Extent of facial nerve schwannoma

	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	Total
IAC		■		■		■		■			■		■			■			7
Labyrinthine		■			■								■	■		■	■	■	7
GG			■							■			■	■	■	■	■	■	8
Tympanic			■						■	■		■		■	■	■	■	■	9
Mastoid	■		■				■		■	■		■		■	■		■	■	10
Extra mastoid												■							1

IAC internal auditory canal, *GG* geniculate ganglion

and recovered to grade III at postoperative 12 months. Tumor removal was not attempted and only decompression was performed in one patient (patient #11). In another patient (case #9) with FNS in the tympanomastoid portion and normal facial function, fascicle preservation surgery was attempted but only decompression could be performed because a cleavage plane could not be found.

Thirteen of 18 patients (72.2%) had preoperative facial palsy of various severity, and gross or near total (case #2) removal of the tumor with FN reconstruction was performed. All patients showed a facial function H-B grade VI immediately after surgery, and 9 of 13 cases (69.2%) recovered to grade III one year after the operation. However, facial function did not improve to better than H-B grade IV in four patients (30.8%) at last follow-up (Table 1).

Outcomes of FN reconstruction

FN was reconstructed in 12 cases using cable graft (10/12, 83.3%), direct anastomosis (1/12, 8.3%), or facial-hypoglossal nerve transfer (1/12, 8.3%). In a case with long-standing facial paralysis (case #10), reconstruction of the FN was not performed and a static facial reanimation was performed at 13 months after the primary surgery. Cable grafts were performed mostly using the sural nerve (9/10, 90.0%) and GAN was used in one case (case #1).

Outcomes were analyzed in 12 patients who underwent nerve reconstruction after tumor removal. Tumor size and duration of preoperative facial paralysis did not significantly affect the final H-B grade after surgery (Fig. 2). However, statistical significance was observed in the final H-B grade after surgery based on the preoperative facial function ($p = 0.046$). In the group with a good facial function (H-B grade II–III) before surgery, the chance of final good facial function (H-B grade III) was high (7/7, 100%). Conversely, patients with preoperative H-B grade of IV or worse had only a 40% (2/5) chance of improving to grade III after surgery (Table 3). There were no complications other than facial palsy after surgery in all patients.

Discussion

The most important goal in the treatment of FNS is the complete removal of the tumor while preserving FN function. To achieve this goal, choosing the appropriate treatment method and optimal timing is important. However, because the incidence is very low and the severity of symptoms varies depending on the location and tumor size, the

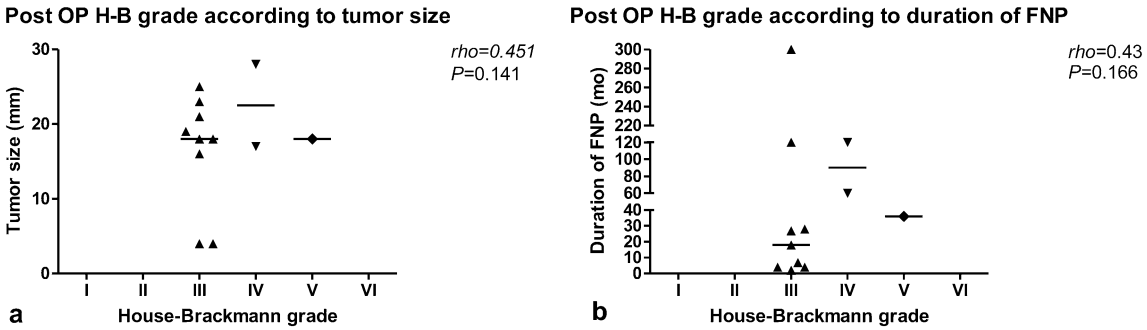

Fig. 2 Postoperative House-Brackmann (H-B) grade based on preoperative tumor size and duration of facial nerve paralysis (FNP). When patients were followed for more than 1 year, preoperative tumor size (**a**) and the duration of facial palsy (**b**) did not affect final H-B grade after surgery

Table 3 Comparison of postoperative H-B grade based on preoperative H-B grade

Variable	Postoperative H-B grade		*p* value
	I–III	IV–VI	
Preoperative H-B grade			0.046
I–III	7	0	
IV–VI	2	3	

H-B grade House-Brackmann grade

determination of appropriate treatment methods and timing for FNS remains problematic [4].

Generally, there are three strategies for managing FNS. The first option is observation and regular check-up using MRI, which may be a good choice if the tumor size is small and symptoms are not present, however, the growth rate of the tumor should be monitored. Various growth rates of FNS of 0.85–1.4 mm per year have been reported, however, growth up to 6 mm per year has occurred [4, 20]. Yang et al. [21] found that larger tumors may grow more rapidly. The authors compared the growth rate of small (< 10 mm) and large (≥ 10 mm) tumors after a follow-up period of 6.4 years. Tumor growth was observed in 72.7% of patients in the large tumor group and 10% in the small tumor group. The authors suggested the 'wait-and-see' policy should be reconsidered for large tumors even in patients with favorable FN function for preservation of remaining hearing, vestibular function, and prevention of intracranial manifestation [3, 17]. The second option is SRS. In a meta-analysis, the tumor control rate was 93.3% after 2 years of follow-up after SRS, however, 12.8% of cases showed deteriorated FN function and 36.7% of cases had worsening of hearing [10]. In another systematic review, SRS resulted in clinical stabilization in most cases (90.7%) [22]. Therefore, SRS may be a good management option for FNS although additional studies with long-term follow-up results are required. Lastly, the surgical management provides complete tumor removal with variable FN morbidity or prolonged preservation of FN function. The facial function, tumor size, location, growth rate, and patient age should all be considered for determining the appropriate treatment modality.

For FNS identified incidentally during surgery for VS, the management option depends on the preoperative severity of facial palsy. If the patient has a poor preoperative facial function (≥ H-B grade III), complete tumor resection and nerve reconstruction should be considered. Subtotal removal or only decompression is recommended for patients with good preoperative FN function (≤ H-B grade II) [17]. Preoperative suspicion of FNS is very important, and the treatment option and possible morbidity of the procedure should be included in the preoperative counseling [23].

The duration of preoperative FNP and final outcomes have been analyzed in several studies [14, 15]. Based on the results, a preoperative deficit duration of less than 1 year is appropriate. In the present study, the duration of facial palsy before surgery did not correlate with preoperative H-B grade or final facial function. Even if the duration of FNP is long enough, recovery to H-B grade III was possible (cases #16 and #17). Tumor size also showed no significant correlation with the final facial function after surgery. However, as mentioned earlier, if the tumor size is large (≥ 10 mm), short-term follow-up is considered more necessary than in cases with small tumor size.

The preoperative facial function is an important factor for determining the timing of surgery. Although in some reports, early surgery was recommended for young patients even when they have a normal facial function [24]. The best FN outcome after grafting or direct anastomosis was reportedly H-B grade III [25]. Therefore, surgical management can be justified only if the facial function is grade III or worse. Among 12 patients in the present study who underwent FN reconstruction, H-B grade remained grade III after surgery in patients with preoperative grade III or better (7/7, 100%). However, patients with grade IV or worse showed improved

facial function up to grade III after surgery in only 2 of 5 patients (40%). Therefore, if the patient's facial function is H-B grade III or better at the time of surgery, obtaining postoperative H-B grade III is very likely. The sustained axonal degeneration precludes adequate stimulation of the facial expression muscle, and subsequent atrophy of the facial muscle is probably the main reason the outcome is unfavorable, and eventually, a muscle transfer surgery is necessary in patients with preoperative H-B grade IV or worse [26, 27].

In summary, due to the small number of patient cases, the effects of multiple factors, such as tumor size and duration of FNP or degree of FNP before surgery, on the final surgical outcomes were not clearly demonstrated. The results of the present study indicate that preoperative H-B grade had a significant effect on the final outcomes. Therefore, regardless of the tumor size and duration of FNP, if the patient with FNS presents with or shows the progression to H-B grade III or worse, surgery should be strongly considered.

Conclusion

Preserving the facial function after surgical removal of the tumor is difficult in FNS cases. Therefore, selecting the appropriate timing of surgical management is important. Based on our experience, facial function H-B grade III is the best time for surgical intervention regardless of the preoperative tumor size or duration of facial paralysis.

Author contributions YSC and Y-SC designed the research; YSC and JEC collected data; YSC and JHL analyzed data; YSC wrote the main paper; Y-SC provided critical revision, discussed the results and implications, and commented on the manuscript at all stages.

Funding The authors have no funding and financial relationships.

Declarations

Conflict of interest All the authors declare that they have no conflict of interest.

Ethical approval This article does not contain any animal studies conducted by the authors. The study was conducted in accordance with the guidelines of the Helsinki Declaration and its subsequent amendments.

Informed consent Informed consent was not needed; it is a retrospective study.

References

1. Lahlou G, Nguyen Y, Russo FY, Ferrary E, Sterkers O, Bernardeschi D (2016) Intratemporal facial nerve schwannoma: clinical presentation and management. Eur Arch Otorhinolaryngol 273:3497–3504
2. Lipkin AF, Coker NJ, Jenkins HA, Alford BR (1987) Intracranial and intratemporal facial neuroma. Otolaryngol Head Neck Surg 96:71–79
3. Yetiser S (2018) Large intratemporal facial nerve schwannoma without facial palsy: surgical strategy of tumor removal and functional reconstruction. J Neurol Surg A Cent Eur Neurosurg 79:528–532
4. Perez R, Chen JM, Nedzelski JM (2005) Intratemporal facial nerve schwannoma: a management dilemma. Otol Neurotol 26:121–126
5. Van Den Abbeele T, Viala P, Francois M, Narcy P (1999) Facial neuromas in children: delayed or immediate surgery? Am J Otol 20:253–256
6. Angeli SI, Brackmann DE (1997) Is surgical excision of facial nerve schwannomas always indicated? Otolaryngol Head Neck Surg 117:S144-147
7. Liu L, Xiang D, Li Y, Sun J (2015) Clinical characteristics and outcomes of intratemporal facial nerve neurofibromas. Am J Otolaryngol 36:565–567
8. Lee WS, Kim J (2011) Revised surgical strategy to preserve facial function after resection of facial nerve schwannoma. Otol Neurotol 32:1548–1553
9. Zabel A, Debus J, Thilmann C, Schlegel W, Wannenmacher M (2001) Management of benign cranial nonacoustic schwannomas by fractionated stereotactic radiotherapy. Int J Cancer 96:356–362
10. McRackan TR, Wilkinson EP, Brackmann DE, Slattery WH (2015) Stereotactic radiosurgery for facial nerve schwannomas: meta-analysis and clinical review. Otol Neurotol 36:393–398
11. O'Donoghue GM, Brackmann DE, House JW, Jackler RK (1989) Neuromas of the facial nerve. Am J Otol 10:49–54
12. Pillsbury HC, Price HC, Gardiner LJ (1983) Primary tumors of the facial nerve: diagnosis and management. Laryngoscope 93:1045–1048
13. Horn KL, Crumley RL, Schindler RA (1981) Facial neurilemmomas. Laryngoscope 91:1326–1331
14. Falcioni M, Taibah A, Russo A, Piccirillo E, Sanna M (2003) Facial nerve grafting. Otol Neurotol 24:486–489
15. Ozmen OA, Falcioni M, Lauda L, Sanna M (2011) Outcomes of facial nerve grafting in 155 cases: predictive value of history and preoperative function. Otol Neurotol 32:1341–1346
16. McMenomey SO, Glasscock ME 3rd, Minor LB, Jackson CG, Strasnick B (1994) Facial nerve neuromas presenting as acoustic tumors. Am J Otol 15:307–312
17. Bacciu A, Medina M, Ben Ammar M, D'Orazio F, Di Lella F, Russo A, Magnan J, Sanna M (2014) Intraoperatively diagnosed cerebellopontine angle facial nerve schwannoma: how to deal with it. Ann Otol Rhinol Laryngol 123:647–653
18. Eshraghi AA, Oker N, Ocak E, Verillaud B, Babcock T, Camous D, Kravietz A, Morcos J, Herman P, Kania R (2019) Management of facial nerve schwannoma: a multicenter study of 50 cases. J Neurol Surg B Skull Base 80:352–356
19. House JW, Brackmann DE (1985) Facial nerve grading system. Otolaryngol Head Neck Surg 93:146–147
20. Wilkinson EP, Hoa M, Slattery WH 3rd, Fayad JN, Friedman RA, Schwartz MS, Brackmann DE (2011) Evolution in the management of facial nerve schwannoma. Laryngoscope 121:2065–2074
21. Yang W, Zhao J, Han Y, Yang H, Xing J, Zhang Y, Wang Y, Liu H (2015) Long-term outcomes of facial nerve schwannomas with favorable facial nerve function: tumor growth rate is correlated with initial tumor size. Am J Otolaryngol 36:163–165
22. Comps JN, Tuleasca C, Goncalves-Matoso B, Schiappacasse L, Marguet M, Levivier M (2018) Upfront Gamma Knife surgery for facial nerve schwannomas: retrospective case series analysis and systematic review. Acta Neurochir (Wien) 160:987–996
23. Kertesz TR, Shelton C, Wiggins RH, Salzman KL, Glastonbury CM, Harnsberger R (2001) Intratemporal facial nerve neuroma:

anatomical location and radiological features. Laryngoscope 111:1250–1256
24. Saleh E, Achilli V, Naguib M, Taibah AK, Russo A, Sanna M, Mazzoni A (1995) Facial nerve neuromas: diagnosis and management. Am J Otol 16:521–526
25. Bozorg Grayeli A, Mosnier I, Julien N, El Garem H, Bouccara D, Sterkers O (2005) Long-term functional outcome in facial nerve graft by fibrin glue in the temporal bone and cerebellopontine angle. Eur Arch Otorhinolaryngol 262:404–407
26. Spector JG (1985) Mimetic surgery for the paralyzed face. Laryngoscope 95:1494–1522
27. Langworth EP, Taverner D (1963) The prognosis in facial palsy. Brain 86:465–480