Facial Nerve Neuroma: Surgical Concept and Functional Results

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ABSTRACT

This study reviewed the management and outcomes of 11 facial nerve neuromas treated in our institution during the past two decades with particular emphasis on surgical concepts and functional outcomes. All patients underwent complete surgical resection of their tumor. Eight patients (73%) were followed on an outpatient basis. A retrospective chart review for pre- and postoperative clinical and radiological data was performed. All facial neuromas were multisegment tumors. All segments of the facial nerve were represented, but 54% involved the geniculate ganglion and 45% involved the labyrinthine or tympanic portions of the nerve, or both. Depending on the extent of sensorineural hearing loss, surgical removal was performed through the middle cranial fossa or translabyrinthine approach. To obtain adequate nerve reconstruction, we combined intra- and extracranial approaches (e.g., the transmastoidal and transtemporal routes). Regardless of the type of nerve reconstruction, the best recovery achieved was moderate facial weakness (House-Brackmann Grade III) in 75% of the patients, even in a patient who was Grade IV preoperatively. The choice of treatment for facial neuromas and surgical approach depends on the extent of tumor, grade of facial palsy, and hearing function. When facial palsy is present, complete resection is clearly indicated. In patients without facial dysfunction, a conservative strategy consisting of clinical and radiological observation should be considered as a treatment option.

KEYWORDS: Facial nerve neuroma, facial nerve tumor, facial schwannoma, facial nerve surgery, facial nerve function

Facial nerve neuromas are rare, composing far fewer than 1% of all intrapetrous mass lesions.¹ The incidence of intratemporal facial neuromas was 0.8% in a cadaveric study, although this figure is higher than the rate of clinical presentation.² To date, about 500 cases of facial neuromas have been reported.³ Facial neuromas are slow-growing tumors that arise from the Schwann cell sheath of the facial nerve. Most originate from the intratemporal portion of the facial nerve.^{3,4} The geniculate ganglion as well as the labyrinthine and tympanic portions of the nerve are the segments most frequently affected,^{3,6,7} and multiple-segment tumors are more common than single-segment facial neuromas.^{3,6}

Clinical symptoms depend on the location of the tumor. Intrameatal lesions usually cause vestibulo-cochlear dysfunction first. Tumors of the geniculate ganglion and tympanic and mastoidal segments produce facial dysfunction, ranging from spasm to palsy. Facial neuromas of the tympanic portion of the nerve can cause conductive hearing loss, and extratemporal tumors show a palpable mass within the parotid gland.³⁻⁸

The advent of magnetic resonance imaging (MRI) has significantly affected the diagnosis and management of facial neuromas by allowing earlier diagnosis, by delineating the extent of the tumor more accurately than was previously possible, and by allowing their differentiation from acoustic neuromas. However, the treatment of facial neuromas is challenging, especially related to functional deficits, and is therefore controversial.^{9,10} We present the clinical findings, surgical management, and functional outcomes of a relatively large series of patients with facial neuromas from a single institution.

MATERIAL AND METHODS

In this retrospective study, 11 patients (6 females, 5 males; mean age, 53 years; range, 30 to 81) with a facial neuroma who were treated at the Department of Otorhinolaryngology of the Teaching Hospital Fulda of the Philipps-University Marburg between

1989 and 2004 were analyzed. The patients' records were reviewed for clinical findings, neuroradiological features, surgical outcomes, postoperative results, and follow-up. Preoperative computed tomography (CT) scans and MRIs were evaluated to determine the size and location of all tumors and surrounding bony erosions. Nine patients had an intratemporal facial neuroma; two were located extratemporally. All tumors were excised completely with histologically controlled free margins. One tumor was diagnosed as a neurofibroma, indicating neurofibromatosis type II. Nine patients were available for follow-up on an outpatient basis (median, 34.2 months; range, 1 to 67 months).

RESULTS

The most common clinical findings in nine patients with an intratemporal facial neuroma were facial palsy (6 cases, 67%) followed by sensorineural hearing loss (3 cases, 33%) and vertigo (2 cases, 22%; Table 1). The two extratemporal facial neuromas presented with a parotid mass without facial nerve dysfunction. All intratemporal facial neuromas were multisegment tumors. The segments most commonly involved were the geniculate ganglion (54%) and labyrinthine and tympanic segments (45% each, Table 2). Only three tumors (33%) extended into the middle cranial fossa extension; one extended into the stylomastoid foramen (Case 5).

Complete tumor resection was achieved in all patients by total excision of the affected nerve segments. The anticipated location of the facial neuroma and the possibility of preserving hearing dictated the surgical approach adopted in each case. Extratemporal tumors were removed by a parotidectomy. In general, the translabyrinthine approach was used when the facial neuroma was located cranial to the geniculate ganglion and severe sensorineural hearing loss was already present (Cases 5, 7, 8 and Fig. 1). Patient 5 became deaf after an earlier cholesteatoma operation. Tumors arising distal to the geniculate ganglion were resected via

Table 1 Patient and Tumor Characteristics, Surgical Management, and Results in 11 Cases of Facial Nerve Neuromas

Pt.	Age (yr)	Sex	Leading Symptom	Facial Nerve Segments Involved	Surgical Approach	Facial Nerve Repair	Follow-up (months)	Facial Nerve Function (House-Brackmann Grade ¹¹)	
								Preop	Postop
1	30	M	Conductive HL	GG, TYMP, MAS	TM	GAN	-	I	-
2	31	M	FP	GG, LAB	TT	GAN	30	IV	III
3	61	M	FP	MAS, TYMP, GG	TM	GAN	12	IV	IV
4	71	F	Parotid mass	EXT	Cervical	SN	63	I	III
5	53	M	FP	MAS, TYMP, GG, LAB	Cervical + TL	SN	35	II	III
6	53	F	Parotid mass	EXT	Cervical	GAN	58	I	III
7	54	F	Vertigo, SNHL	LAB, IAC, CPA	TL	GAN	67	I	III
8	46	F	Vertigo, SNHL	IAC, CPA	TL	HGN	-	I	-
9	81	F	FP	GG, TYMP, LAB	TM	HGN	16	IV	IV
10	34	M	FP	IAC, CPA	TT	SN	26	IV	III
11	65	F	FP, SNHL	GG, LAB, TYMP	TT + TM	SN	1	III	-

HL, hearing loss; GG, geniculate ganglion; TYMP, tympanic segment; MAS, mastoid segment; TM, transmastoid; GAN, greater auricular nerve; FP, facial palsy; LAB, labyrinthine segment; TT, transtemporal; HGN, hypoglossal nerve; EXT, extratemporal; SN, sural nerve; TL, translabyrinthine; SNHL, sensorineural hearing loss; IAC, internal auditory canal; CPA, cerebellopontine angle.

transmastoid exploration (Cases 1, 3, 9, 11). The transtemporal approach was performed in two patients to preserve their hearing (Cases 2, 10 and Fig. 2). In one woman transmastoid and middle cranial fossa procedure were combined (Case 11). No surgical complications occurred, and no recurrent tumors were observed.

The preferred method of facial nerve repair was interposition of a greater auricular nerve or sural nerve cable graft, depending on the length needed. A facial nerve-hypoglossal nerve anastomosis was performed in two cases in which the central stump of the facial nerve was too short to be sutured. All

patients who could be followed recovered partial facial function, achieving moderate facial weakness (House-Brackmann¹¹ Grade III) in 75%. A gold lid implant was used for facial nerve rehabilitation in all patients.

DISCUSSION

About 5% of peripheral facial nerve palsies are caused by tumors.² Of these, facial neuromas compose only a small fraction.^{5,8} These slow-growing tumors probably arise from the sensory fibers of the facial nerve.⁸

A review of the literature summarized 467 cases published up to 2002.³ Among the articles reviewed, one publication described seven cases of facial neuromas in children.¹² Our study confirms the finding that facial neuromas can occur at any age with no significant gender predilection.^{3,6} The results of our series also underscore previous reports indicating that clinical facial weakness (63.2% of cases) and hearing loss (50.6% of cases) are the predominant presenting symptoms.⁶ Facial palsy

Table 2 Location of Facial Nerve Neuroma by Segment

Anatomical Segment	Number of Cases (%)
Cerebellopontine angle	3 (27%)
Inner auditory canal	3 (27%)
Labyrinthine segment	5 (45%)
Geniculate ganglion	6 (54%)
Tympanic segment	5 (45%)
Mastoid segment	3 (27%)
Extratemporal segment	2 (18%)

Figure 1 Case 7. (A) Gadolinium-enhanced T1-weighted axial MRI showing a left-sided tumor mass from the labyrinthine facial segment through the inner ear canal to the cerebellopontine angle (circle). Clinically, the patient complained of complete hearing loss without signs of facial palsy. The tumor was resected through a translabyrinthine approach. The facial nerve was reconstructed with a greater auricular nerve graft. (B) Postoperative gadolinium-enhanced T1-weighted axial MRI.

was the primary symptom in our patients (67%), while sensorineural hearing loss occurred in 33% and vertigo in 22%. Both patients with an extra-temporal facial neuroma had a parotid mass. The interval between the onset of symptoms and diagnosis was short (up to 1 year) when facial dysfunction was present but much longer with hearing loss (up to 5 years).

Any segment of the facial nerve may be involved with a neuroma. Since the introduction of MRI for the evaluation of facial neuroma, the geniculate ganglion and labyrinthine segments have been identified as the segments most likely to be

involved. Their involvement can be attributed to the major structural reorganization of the nerve that occurs in these regions.^{3,6} The most frequently affected segments in our facial neuroma series were the geniculate ganglion (54%) and labyrinthine and tympanic segment (45%). These findings are about comparable to other reports in which 43.5% of the tumors involved the labyrinthine/geniculate segment, 42.8% involved the tympanic, 36.7% involved the mastoidal segment, and 24.3% involved the internal auditory canal (IAC).³ The sensitivity of MRI also may account for a decrease in the misdiagnosis of facial neuroma. After MRI

Figure 2 Case 2. (A) Axial CT showing soft tissue density mass (arrow) at the right geniculate ganglion with surrounding bone erosion. (B) T1-weighted axial MRI showing the gadolinium-enhanced lesion at the geniculate ganglion and tympanic segment. Clinically, the patient presented with facial palsy (Brack-Housemann Grade IV) without hearing loss. To preserve hearing, the tumor was resected via a transtemporal approach. The facial nerve was reconstructed with a greater auricular nerve graft. (C) Postoperative gadolinium-enhanced T1-weighted axial MRI.

became available, the mean number of segments per tumor of 1.89 increased to 2.57.^{5,6} All of the facial neuromas in our patients involved multiple segments of the nerve. In contrast, Kertesz et al⁶ reported that 36.4% of their cases involved only one segment.

The treatment for facial neuromas is surgical excision, but the decision to operate is controversial because facial palsy is inevitable after surgical resection. Liu and Fagan¹⁰ eliminated 10 patients without facial palsy as surgical candidates and followed them for as long as 10 years. The patients showed continued normal facial function, and no tumor growth was evident radiologically in 5 patients. Van Den Abbeele and coworkers¹² reported one child managed conservatively whose facial neuroma remained stable clinically and radiologically after 9 years of follow-up.

Lipkin and associates⁴ reported that the best results from facial nerve grafting are obtained when only moderate dysfunction has developed. In contrast, Symon and colleagues⁵ proposed no delay in surgery because the risk to hearing increased and the outcome of facial nerve reconstruction was best in patients with no facial palsy. No adequate data are available to judge the best timing to perform an operation to obtain the best functional recovery of the facial nerve.

Regardless of the timing of surgery or the type of facial nerve repair, patients can expect no better than an eventual House-Brackmann Grade III palsy.^{5,10} We removed all facial neuromas surgically to perform the anastomosis with the shortest distance possible before further tumor enlargement. In future patients with facial neuroma and no facial dysfunction, we could also imagine recommending observation and serial radiological imaging as the first steps in treatment.

Surgical strategy should be based on the anatomic location and extension of a facial neuroma.¹³ Consequently, the type of hearing disturbance should be the first consideration in choosing an approach. Tumors on the tympanic or mastoidal portion of the nerve can be treated within a transmastoid operation followed by a tympanoplasty. If

the facial neuroma involves segments proximal to the cochleariform process and the patient is deaf, as in our Case 7, we prefer the translabyrinthine route. For adequate nerve reconstruction it can be useful to combine the transmastoidal and transtemporal approaches, especially in large tumors involving multiple segments where an intracranial-intratemporal anastomosis is needed.¹⁴ In patients with a facial neuroma located in the IAC or at the cerebellopontine angle (CPA) without remarkable sensorineural hearing loss, a middle cranial fossa or retrosigmoid approach should be performed, possibly in an interdisciplinary manner with neurosurgeons.^{13,14}

O'Donoghue and coworkers⁸ reported that they were able to preserve at least 50% of the facial nerve intact in 25% of their patients. This has not been our experience, nor that of others.^{10,15} In our hands, surgical excision of the facial neuroma results in complete removal of the involved nerve segments histologically secured by frozen section. To restore the facial nerve we prefer to use the sural or greater auricular nerve as a graft. We found no advantage for any particular type of reconstruction, with the best recovery being moderate weakness (House-Brackmann Grade III).

CONCLUSION

Facial palsy and sensorineural hearing loss are the most common presenting symptoms associated with facial neuromas. High-resolution MRI and CT are mandatory to obtain an accurate diagnosis, to delineate the dimensions of the tumor, and to choose the most appropriate therapeutic strategy. Because most facial neuromas involve the geniculate ganglion and labyrinthine facial nerve segment, surgical removal usually requires a middle cranial fossa or translabyrinthine approach, depending on the degree of sensorineural hearing loss. Early diagnosis may improve the outcome of facial nerve function, because the efficacy of grafting partially depends on the duration of paralysis. In patients with a facial

neuroma without facial dysfunction, careful observation and serial radiological imaging should be considered the first treatment.

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Commentary

Facial nerve neuromas are extremely rare skull base lesions that typically affect the geniculate ganglion and temporal and labyrinthine segments of the facial nerve. Patients present with a variety of symptoms ranging from sensorineural hearing loss to facial palsy. Tumors with multiple segments are common. Surgical resection of these lesions always results in facial palsy. As the authors have pointed out, the maximal improvement that can be expected is House-Brackmann Grade III. Surgical resection should only be undertaken after a detailed discussion with the patient at which informed consent is obtained. Whether surgical palsy is an appropriate trade-off for hearing preservation is a decision that must be made between physician and patient. Although there are no controlled series using gamma knife or fractionated radiosurgery with the Cyberknife, one would expect the results to be similar to those from acoustic neuroma series. However, fractionated radiosurgery to maximize function of the nerve may be more attractive to patients with partial facial nerve palsy.

I thank the authors for sharing this well-written article.

Randall W. Porter, M.D.¹

Commentary

This article reports a relatively large series of facial nerve neuromas treated at a single institution. In all cases, the neuroma was excised and facial nerve reconstruction was performed, typically with a nerve graft. A hypoglossal facial nerve anastomosis was performed only when the proximal facial nerve was unavailable for grafting. Both procedures represent the appropriate standard of care.

The authors discuss the option of observation, which as they note, increasingly is becoming

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the accepted initial treatment for these tumors. At the House Ear Clinic, we recommend observation as long as facial nerve function remains good. However, we discuss the risk of hearing loss with the patient because these tumors can erode into the cochlea and produce sensorineural hearing loss despite continued good facial nerve function.

When facial nerve function begins to deteriorate, surgical resection of the tumor with facial nerve grafting is recommended. The appropriate timing for surgery is a difficult decision. Surgery is usually delayed until the patient has significant facial nerve dysfunction, in the range of a House-Brackmann Grade III or IV. Waiting until the

facial nerve completely degenerates compromises recovery, and the patient is urged not to wait too long to make the decision to undergo surgery.

Our experience is similar to that of the authors. When good facial function is present preoperatively, most patients receiving a facial nerve graft make a Grade III recovery. If the facial nerve has degenerated preoperatively, one usually expects only a Grade IV recovery, which is also expected with a hypoglossal facial anastomosis.

I congratulate the authors for their well-documented series and excellent discussion of this problem.

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