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What is This?
Management Outcomes of Facial Nerve Tumors: Comparative Outcomes with Observation, CyberKnife, and Surgical Management

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Abstract

Objectives. Primary facial nerve tumors (FNTs) present in varying ways. In this study, the authors present their institutional experience with the management of facial nerve tumors, including their recommendations for available therapies such as observation, microsurgical decompression or removal, and stereotactic radiation. They emphasize the auditory and facial nerve function outcomes.

Study Design. Retrospective case review.

Setting. Tertiary referral center.

Subjects and Methods. Retrospective review of all cases of FNT seen at the authors’ tertiary care academic medical center over a 10-year period (2002-2011). The clinical presentation, treatment modality, and outcome parameters of cochlear and facial nerve function were assessed.

Results. Twelve patients were identified. House-Brackmann grades on presentation were 4 grade I, 2 grade II, 2 grade III, 1 grade IV, and 3 grade V, with 2 grade V patients declining to grade VI shortly after presentation. Seven patients presented with serviceable hearing and 4 with nonserviceable hearing. Treatment options/arms included observation with serial clinicoradiological review (2 cases), stereotactic radiation with the CyberKnife (3 cases), wide fallopian canal decompression (3 cases), microsurgical excision and repair (3 cases), and biopsy followed by observation (1 case). At the end of the review period, facial nerve function was stable in 8 patients, improved in 3, and declined in 1, and none had documented worsening of hearing based on American Academy of Otolaryngology—Head and Neck Surgery Foundation classification.

Conclusions. Management of FNT is largely based on the clinicoradiological picture. Each treatment arm is different, but overall auditory and facial function can be maintained.

Keywords

facial nerve, neuroma, schwannoma, fallopian canal, decompression, CyberKnife, radiation

Patients with primary facial nerve tumors (FNTs) typically present with gradual progression and onset of auditory and facial nerve dysfunction and are generally middle aged with a slight female predominance.1-4 Five percent of facial nerve palsies are due to neoplasms. Facial nerve schwannomas (FNSs) are relatively rare benign neoplasms of Schwann cell origin and the most common neoplasm of facial nerve origin, whereas facial nerve neuromas are benign neoplasms of the nerve axon.1,2 Because it is radiologically difficult to distinguish between facial nerve schwannomas and neuromas, we have collectively grouped them as FNTs in this discussion. When FNTs arise within the temporal bone, they present with varying degrees of facial nerve dysfunction and/or hearing loss but may also be entirely asymptomatic and discovered incidentally during temporal bone imaging or procedures for other pathology. When a FNT presents incidentally or with good facial nerve function (HB I-III), it presents a treatment dilemma for the

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neurotologic surgeon because these tumors are notoriously indolent and slow growing.1,3-5

Management options are largely determined by the clinicoradiological picture (especially the degree of facial nerve dysfunction) and include observation, radiotherapy, and surgical intervention.1,3-5 The aim of management is to ensure that the patient has the best facial function possible whether by maintenance or improvement of present function. Observation is commonly employed in patients with good facial nerve function.1,4,6 Facial nerve impairment may progress due to entrapment-induced ischemia of the nerve as well as invasion of the fascicular bundles,5 and intervention becomes a viable option once this ensues. Radiotherapy in the form of CyberKnife or gamma radiation aims to prevent further tumor growth.7,8 Fallopian canal decompression aims to reduce the bony confinements of growth.5 Microsurgical excision and repair involves tumor resection and repair either primarily with an interpositional nerve graft or anastomosis to another nerve (eg, hypoglossal), which typically results in facial nerve outcome of House-Brackmann grade III at best.3,9,10 This article aims to assess the auditory and facial nerve functional outcomes experienced by different therapy arms at our institution.

Methods

After obtaining approval from the University of Miami Institutional Review Board, a retrospective chart review was done identifying all patients with a diagnosis of facial nerve tumors seen at our institution from 2002 to 2011. Twelve patients were identified. Each patient’s symptomatology and duration of symptoms were noted; specifically, facial nerve function based on the House-Brackmann (HB) scale was noted pre- and posttreatment. Patients were audiologically assessed using pure-tone audiometry at presentation and again within several months posttreatment, with subsequent audiograms on a yearly basis if follow-up permitted. Pure-tone audiogram (PTA) and speech discrimination scores (SDS) were carried out using standard audiometric techniques. Pure-tone average was calculated based on the recommendation of the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology—Head and Neck Surgery (AAO-HNS). Hearing was categorized according to the AAO-HNS guidelines in which serviceable hearing was defined as having a PTA better than 50 dB and SDS greater than 50%. The FNTs were assessed initially using high-resolution computerized tomography (HRCT) and contrast-enhanced magnetic resonance imaging (MRI), and those tumors that were observed were followed on a yearly basis with MRI. Management was based predominantly on the clinicoradiological findings. The 4 main tiers of management were observation, radiotherapy, fallopian canal decompression, and microsurgical excision and repair. All cases were operated on by the senior authors (S.I.A. and F.F.T.). CyberKnife radiation was performed at our institution for 2 patients and at an outside facility for another.

Results

Twelve patients were identified. There were 7 men and 5 women. The average age of the patients was 42.4 years (range, 23-62 years). Average follow-up was 35.2 months. All patients had imaging and audiometry performed. House-Brackmann grades on presentation were 4 grade I, 2 grade II, 2 grade III, 1 grade IV, and 3 grade V, and 2 grade V patients declining to grade VI shortly after presentation. Five patients presented with findings on otoscopy, including 3 with middle ear masses and 2 with evidence of cholesteatoma. Seven patients presented with serviceable hearing and 4 with nonserviceable hearing. One patient had no documented audiogram upon presentation. Eleven patients presented with gradual progression of facial nerve symptoms, including twitching, spasm, and palsy. Radiologically, the geniculate ganglion and tympanic segments were most

Figure 1. (A, B) Axial magnetic resonance imaging (MRI) findings in facial nerve tumors (FNTs). Postcontrast axial MRI films demonstrate contrast-enhancing facial nerve masses suggestive of FNT in the perigeniculate region.
commonly involved in 8 and 11 patients, respectively (see Figure 1A, B and Figure 2A, B). Two patients had biopsy-confirmed schwannoma prior to presentation to our clinic, whereas 3 patients had biopsy after presentation: 2 schwannomas and 1 neuroma. Two patients were incidentally diagnosed clinically during otologic surgery for a separate pathology (cholesteatoma), whereas the remainder were initially diagnosed based solely on clinicoradiological findings. Two patients completed the observation arm after 1 defaulted to CyberKnife due to worsening facial nerve function. CyberKnife (CBK) therapy was used in 3 patients. Three patients underwent wide fallopian canal decompression. Microsurgical excision and facial nerve repair (MER) were carried out in 2 patients. One patient had a biopsy followed by serial observation.

Patients’ hearing and facial function were assessed at the time of their last follow-up visit as depicted in Table 1. There was no change or improvement in hearing based on the AAO-HNS classification for 10 patients; 1 had no posttherapy PTA (CBK group) and the other no preoperative PTA (MER group) available. Facial nerve function improved in 1 patient from the CBK arm and 2 in the MER arm, whereas 1 patient had worsening facial nerve function in the CBK arm (HB IV-VI). The remainder had stabilization of their FN function during the follow-up period. Each group’s clinical data are described below and summarized in Tables 1 and 2.

### Observation

Three patients elected to undergo serial clinicoradiological observation (using MRI), but 1 defaulted to CyberKnife due to sudden worsening of the HB score and will be discussed in the CBK group. Neither of the 2 patients completing observation had clinicoradiological changes at the end of the follow-up period. Of note, 1 patient (OB2) presented with a history of idiopathic facial nerve palsy 24 years prior to presentation, which resolved spontaneously. A few months prior

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**Table 1.** Facial Nerve Function, Hearing, and Tumor Extent in 12 Patients with Facial Nerve Tumors

<table>
<thead>
<tr>
<th>Patient</th>
<th>Follow-up, mo</th>
<th>Involved Segment of the Facial Nerve</th>
<th>Facial Function Pretreatment&lt;sup&gt;a&lt;/sup&gt;</th>
<th>Facial Function Posttreatment&lt;sup&gt;a&lt;/sup&gt;</th>
<th>Hearing Posttreatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>OB1</td>
<td>84</td>
<td>Geniculate ganglion to tympanic</td>
<td>II</td>
<td>II</td>
<td>Stable</td>
</tr>
<tr>
<td>OB2</td>
<td>24</td>
<td>Internal auditory canal to mastoid</td>
<td>I</td>
<td>I</td>
<td>Stable</td>
</tr>
<tr>
<td>OB3</td>
<td>48</td>
<td>Tympanic to extratemporal</td>
<td>III</td>
<td>III</td>
<td>Stable</td>
</tr>
<tr>
<td>CBK1</td>
<td>36</td>
<td>Internal auditory canal to tympanic</td>
<td>IV</td>
<td>III</td>
<td>Stable</td>
</tr>
<tr>
<td>CBK2</td>
<td>2</td>
<td>Geniculate ganglion to mastoid</td>
<td>II</td>
<td>VI</td>
<td>No audiogram available</td>
</tr>
<tr>
<td>CBK3</td>
<td>14</td>
<td>Geniculate ganglion to mastoid</td>
<td>I</td>
<td>I</td>
<td>Stable</td>
</tr>
<tr>
<td>FCD1</td>
<td>35</td>
<td>Tympanic to mastoid</td>
<td>I</td>
<td>I</td>
<td>Improved</td>
</tr>
<tr>
<td>FCD2</td>
<td>80</td>
<td>Tympanic to mastoid</td>
<td>I</td>
<td>I</td>
<td>Stable</td>
</tr>
<tr>
<td>FCD3</td>
<td>22</td>
<td>Tympanic to mastoid</td>
<td>III</td>
<td>III</td>
<td>Stable</td>
</tr>
<tr>
<td>MER1</td>
<td>45</td>
<td>Labyrinthine to mastoid</td>
<td>VI</td>
<td>III</td>
<td>No audiogram available</td>
</tr>
<tr>
<td>MER2</td>
<td>53</td>
<td>Labyrinthine to mastoid</td>
<td>V</td>
<td>IV</td>
<td>Stable</td>
</tr>
<tr>
<td>MER3</td>
<td>3</td>
<td>Tympanic to mastoid</td>
<td>VI</td>
<td>VI</td>
<td>Stable</td>
</tr>
</tbody>
</table>

<sup>a</sup>House-Brackmann facial function scale.
to presentation, a sudden increase in facial twitching and eventual facial palsy was noted with pressure changes during an aircraft flight but resolved spontaneously with pressure equalization. A middle ear mass was noted on otoscopy, and a ventilation tube was placed that successfully prevented further occurrences of facial palsy with barometric changes. One patient (OB3) presented with a 2-year history of left facial spasm with a previous biopsy at an outside institution. Examination revealed synkinesis and an HB grade III with an obvious mass eroding through the bony posterior external ear canal wall, which was also seen beneath the tympanic membrane in the mesotympanum. Magnetic resonance imaging and computed tomography (CT) scan confirmed a mass from the tympanic to the infratemporal fossa portion of the facial nerve measuring $2.5 \times 1.7 \times 2.7$ cm. A transmastoid biopsy was done to confirm the diagnosis of schwannoma. There were no subsequent clinical or radiological changes in the 4-year follow-up period. This patient was included in the observation arm as the surgery consisted only of an incisional biopsy, and there was no distinct indication for fallopian canal decompression.

**CyberKnife**

Three patients had CyberKnife treatment, including the 1 default from the observation arm. The indications for CyberKnife were rapid decline in facial nerve function, rapid increase in tumor size, and patient preference (the latter was performed at an outside institution). Mean tumor volume was 2.09 cm$^3$ with mean total dose of 1961.7 cGy.

One patient (CK1) presented with a history of progressively worsening right facial muscle twitching and hearing loss over a period of 18 months and opted for CyberKnife therapy at an outside facility. Another patient (CK2) presented with a history of right facial twitching, spasm, HB grade II, hearing loss, otalgia, and abnormal tearing. An incisional biopsy at an outside institution had confirmed FNS. Otoscopy revealed the presence of a middle ear mass. The patient’s facial function remained stable for 4 years but suddenly declined to HB grade IV with no radiological changes, and CyberKnife therapy was recommended. CK3 presented with more than a 3-year history of hearing loss, no facial nerve symptoms, and a red mass on otoscopy. This patient had MRI-documented doubling of the tumor size 3 years prior to presentation. Facial nerve function outcomes varied in the CBK group, with 1 patient improving, 1 declining, and 1 maintaining stable facial nerve function. Hearing was stable in 2 patients, and no posttherapy PTA was documented for 1 patient (patient did not present for audiogram).

**Microsurgical Fallopian Canal Decompression**

Three patients were managed with wide fallopian canal decompression (FCD). Two patients presented with chronic otitis media and normal facial nerve function, and both had their tumor discovered incidentally during routine cholesteatoma surgery. The diagnoses were based on the clinical picture. Both of these patients had their FNT decompressed during cholesteatoma surgery, an intraoperative decision made by the primary surgeon.

The third case presented with HB grade III, and diagnostic imaging confirmed an enhancing lesion along the course of the facial nerve. An incisional biopsy confirmed the diagnosis of facial nerve schwannoma. This patient elected to undergo FCD given the possibility of further decline in facial nerve function, which was already HB III.

There were no changes in the facial nerve function of any of the 3 patients undergoing FCD at the end of the review. Postoperatively, hearing thresholds remained stable or improved in all 3 patients.

**Microsurgical Resection and Repair**

All 3 patients in our microsurgical resection and repair group presented with poor facial nerve function (grade V), and 2 patients declined to grade VI shortly after presentation. All were managed with resection of the FNT and repair with interpositional nerve grafts. Frozen-section pathology reported the presence of 1 schwannoma (MA1) and 1 facial nerve neuroma (MA3). Facial nerve function improved in 2 patients and was unchanged in 1 patient at the end of the review period. There were no documented audiometric changes, although MER1 had no documented pretherapy audiogram for comparison.

**Discussion**

It is difficult to draw conclusions regarding the surgical treatment of FNT as they only comprise 0.15% to 0.8% of...
all intratumoral tumors. Opinions vary as to the management of facial nerve tumors. Most authors seem to agree that when patients present with minimal symptoms (ie, good HB score [I-II]), the ideal management is observation.\textsuperscript{1,3,4,9,11} This is advocated because of the slow growth of the tumor and the distinct possibility that there will be no worsening of present symptoms for decades.\textsuperscript{1,3} Also, interventional therapies are not without risks of further facial nerve and auditory function impairment.\textsuperscript{7,8} In our series, 2 patients remained clinicoradiologically unchanged over the average review period of 54 months, which is consistent with findings of Liu et al,\textsuperscript{3} who reviewed 10 patients managed conservatively with FNT and noted no decline in facial nerve function in a 10-year review. Observation therefore remains a suitable option in patients with good facial nerve function (HB I-II) with a radiologically stable picture.

If patients have a worsening clinicoradiological picture (HB grade II-IV, evidence of radiological FNT growth, or impending complications such as erosion into the labyrinthine capsule), treatments such as FCD, radiation, or MER are options that should be carefully considered based on the particular patient’s situation.\textsuperscript{5,7,8} The use of radiation to stabilize or reverse tumor growth has been documented.\textsuperscript{7,8} Advantages include avoidance of major surgery and the period of delay in HB improvement (seen after nerve repair). Another advantage may be seen in rapidly growing tumors in patients with good facial function (HB II) who are poor surgical candidates. Radiation, including gamma knife (GK) therapy, has been documented to stabilize tumor growth and facial nerve function.\textsuperscript{7,8,12} The use of CyberKnife in FNT management is a relatively new area and has not been well documented in the literature. It involves the use of a linear accelerator with 0.5-mm accuracy similar to GK. The advantages, however, include no need for a rigid frame bolted to the skull, the ability to divide therapy into several sessions to preserve vital adjacent structures (eg, CN VIII, cochlea), and real-time imaging guidance tracking for robotic adjustment to movement. This series represents one of few in the literature to use CyberKnife in the management of FNT, although our results do not discern any posttreatment trends given our small sample size and disparate outcomes. Radiation offers an alternative option to patients with early facial dysfunction (HB II-III) with a documented worsening of the clinicoradiological picture. It has a small but notable risk of worsening auditory and facial function, which should be included in any management discussion.

Fallopian canal decompression is an alternative option, especially in patients with large expanding tumors, and may delay the onset or progression of facial nerve paresis/palsy.\textsuperscript{5,13} Fallopian canal decompression does not reduce tumor growth but rather reduces the bony confinement and subsequent nerve ischemia/paresis. Thus, CyberKnife or microsurgical excision would be more appropriately considered if there is impending labyrinthine erosion. In 1997, Angeli and Brackmann\textsuperscript{3} first described FCD in patients with facial nerve tumors and reported that all patients experienced improvement or stability of facial nerve function postoperatively. Disadvantages include the need for major surgery and the subsequent risk of auditory, facial nerve, and vestibular dysfunction, but FCD avoids the need for a prolonged delay in facial nerve recovery, as seen in nerve repair as well as the risks associated with radiotherapy.\textsuperscript{3,5,9} It is a viable option for varying HB grades with radiological evidence of tumor growth and FNT discovered incidentally during surgery for other otologic pathology. It offers patients the option of maintenance of facial nerve function if they have limited dysfunction or do not wish to undertake excision. We recommend decompression of all of the involved facial nerve segments based on preoperative MRI enhancement and visual intraoperative inspection. The surgeon needs to be ready to extend the decompression to the perigeniculate and labyrinthine segments if needed; the middle fossa craniotomy approach is best for patients with serviceable hearing, but a trans-mastoid/trans-labyrinthine approach to the labyrinthine segment is appropriate for patients with nonserviceable hearing. We limited our FCD to a trans-mastoid approach given that in 2 patients, there was no radiological or intraoperative evidence of tumor extent beyond the tympanic and mastoid segments. Our experience was consistent with published literature, demonstrating stable facial nerve function and hearing outcomes with decompression.

Microsurgical excision and facial nerve repair is the only option involving tumor resection. Partial excision of FNT in patients with good facial nerve function is not universally recommended due to the risk of nerve injury. Microsurgical excision and facial nerve repair is usually recommended for patients with poor HB grade IV to V and in whom tumor compression on vital structures (eg, brainstem or labyrinthine erosion) is evident.\textsuperscript{3,4,9,10} Postoperative facial nerve paralysis for 6 to 18 months is expected followed by improvement to a HB grade III at best.\textsuperscript{3,4,9-11,13} Commonly used repair options include primary repair, interpositional nerve graft (greater auricular and sural nerves are commonly used), and hypoglossal-facial anastomosis. The earliest and best results are seen with primary repair.\textsuperscript{3,4,9,10} Shirazi et al\textsuperscript{10} advocated complete surgical resection of the tumor when facial palsy exists and presented a series of 15 patients with various methods for nerve repair following tumor excision. None of the 15 patients had facial nerve function better than HB grade III, which is consistent with other published literature for MER. Our results were consistent with the literature, demonstrating an improvement in 2 of 3 patients after excision and repair but none with better than HB grade III.

Conclusion
Management modalities for FNT vary and are largely based on the clinicoradiological presentation. There are many management options available, including observation, radiation, FCD, and MER. Because the disease progress tends to be slow, observation with a “scan-and-wait” approach is an acceptable modality of treatment. Once the clinicoradiological
picture worsens (especially HB grade or tumor size), interventional management should be considered in the form of radiotherapy, wide fallopian canal decompression, or microsurgical excision and repair. Interventional modalities of surgery and radiotherapy are not without risks and routinely do not improve facial nerve function above HB II to III at best. In this series, we have reviewed our experience with the varying treatment options, including the use of CBK to treat FNT, which add to the small number of such cases reported in the literature. Our cases demonstrate that satisfactory facial and auditory outcomes are possible via any treatment arm. Our current treatment algorithm relies on observation with serial MRI for FNT with no or limited facial nerve dysfunction, whereas deterioration of facial nerve function or hearing, rapid growth, or impending labyrinthine erosion or other complications warrant consideration of other modalities. Radiation or FCD is most often employed for tumors with HB grade II to IV facial nerve function. Microsurgical excision and facial nerve repair is generally delayed until HB grade IV to VI because at this point, there is essentially only room for improvement. It is of paramount importance to offer patients the full range of options, so that they may play an active role in guiding treatment, because there is no gold standard for this rare neurotologic entity.

Author Contributions

Guyan A. Channer, data acquisition, drafting of initial article, article editing, final approval for article to be published; Björn Herman, data acquisition, drafting of initial article, article editing, final approval for article to be published; Fred F. Telischi, article design, data analysis, article revision, final approval for finished article; Daniel Zeitler, data acquisition, drafting of initial article, final approval for article to be published; Simon I. Angeli, design of article, data analysis and interpretation, article editing, final approval for finished article.

Disclosures

Competing interests: Simon Angeli, Medtronic, research grant (research subject is not related to this manuscript).

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