Intratemporal Hemangiomas Involving the Facial Nerve: Diagnosis and Management

Abstract—Intratemporal vascular tumors involving the facial nerve are rare benign lesions. Because of their variable clinical features, they are often misdiagnosed preoperatively. This study presents a series of 21 patients with such lesions managed from 1977 to 1994. Facial nerve dysfunction was the most common complaint, present in 60% of the cases, followed by hearing loss, present in 40% of cases. High-resolution computed tomography, magnetic resonance imaging with gadolinium, and a high index of clinical suspicion is required for preoperative diagnosis of these lesions. Early surgical resection of these tumors permits acceptable return of facial nerve function in many patients. (*Skull Base Surgery*, 5(4):227–232, 1995)

Intratemporal hemangiomas are rare benign tumors, commonly arising either at the geniculate ganglion (GG) or within internal auditory canal (IAC). They are regarded as vascular hamartomas or hamartoblastomas rather than true neoplasms and are often indistinguishable from vascular malformation.¹

Clinically, these lesion when located in the IAC imitate the symptomatology of acoustic neuromas with hearing loss as the predominant feature. Facial nerve dysfunction is more common when the tumors are situated at the GG, and many patients present as having recurrent Bell's palsy.² Thus, the relative rarity and their variable clinical presentation along with paucity of clinical experience with these lesion often leads to delayed diagnosis.³

The aim of this study is to present our experience in a series of 21 intratemporal vascular tumors (hemangiomas and vascular malformations) involving the facial nerve operated on by the senior authors (A.M., M.S.), emphasizing their variable clinical presentations and the relevant surgical management.

MATERIAL AND METHODS

The medical records of 21 patients (13 male, 8 female) of intratemporal vascular tumor involving the facial nerve managed from 1977 to 1994 were reviewed. Seven of these patients have been previously reported.^{4–6} Their ages ranged from 8 to 66 years, with an average age of 45 years. Twenty of these cases were histologically proved (Table 1); in one case the diagnosis of hemangioma was likely, but could not be confirmed histologically and is excluded from the present study.

CLINICAL FEATURES

In this series the majority of patients had more than one clinical symptom. Except for one case with IAC lesion presenting with the sole complaint of tinnitus, in all the remaining cases hearing loss was the first clinical symptom, whereas facial nerve dysfunction was the pre-

Table 1. Histopathology of Intratemporal Vascular Tumors Involving the Facial Nerve

<i>Histopathology</i>	<i>Cases</i>
Hemangioma cavernous	6
Angiofibroma	6
Ossifying hemangioma	3
Hemangioma arteriovenous type	2
Vascular malformation	1
Hemangiopericytoma	1
Hemolymphangioma	1
Total	20

dominant symptom in cases with a GG lesion. Facial nerve dysfunction occurred either in the form of hemifacial spasm, facial paresis, or paralysis in 12 cases (60%). The symptom of hearing loss was present in eight cases (40%), tinnitus in seven cases (35%), and otorrhea in only one case (5%).

DIAGNOSIS

Preoperative diagnostic evaluation included audiometric and radiologic investigation. The preoperative audiometric test revealed normal hearing in eight patients (40%), total deafness in seven cases (35%), moderate sensorineural hearing loss (SNHL) in two cases (10%), and conductive deafness in one case (5%). Audiologic data were not available for two patients (10%). The preoperative and postoperative hearing status is summarized in Table 2.

Apart from two early cases, all the remaining cases underwent either a computed tomography (CT) scan or magnetic resonance imaging (MRI) evaluation. High-resolution CT scan and MRI with gadolinium are complementary and were found to be the most essential tools for preoperative diagnosis of these tumors. However, despite advanced neuroradiologic investigation, the preoperative diagnosis of vascular tumor involving the facial nerve was made in only three cases (15%), facial nerve schwannoma in five cases (25%), acoustic neuroma in nine cases (45%); all the tumors were located in IAC, and in the remaining one case (5%) petrous bone cholesteatoma was

suspected. Preoperative clinical impression was not available from the medical records of two patients (10%).

TREATMENT

The surgical approach was decided according to the location of the tumor, preoperative hearing status, and tumor size. The technical details of various surgical procedures used are as described by Sanna et al.⁷ Middle fossa alone or combined with transmastoid approach was used in patients with good hearing and in whom total facial nerve exposure was necessary. The transmastoid approach was used in cases with tumors of tympanic and vertical portion of the facial nerve. The transmastoid extralabyrinthine approach⁸ was used in cases with tumor extending into the GG and the lateral part of the labyrinthine portion of the facial nerve. In cases with preoperative unserviceable hearing or total hearing loss, translabyrinthine approach was used. Table 3 summarizes the different approaches used in the management of these patients.

The majority of tumors were less than 1 cm in size, with a range of 0.3 to 3 cm. Nine tumors were centered in the IAC, nine in the GG, and two in the middle ear. The facial nerve was anatomically preserved in five cases. In 12 cases facial nerve was repaired using either sural or greater auricular nerve grafts and facial-hypoglossal anastomosis was required in one case. In the remaining two cases, facial nerve repair was not attempted, as both of them had a 20-year history of complete facial palsy. One year postoperative results were available in 12 cases. Two patients were lost to follow-up and three cases managed recently in the past 6 months did not have a long enough follow-up. Facial nerve results are detailed in Table 4.

CASE ILLUSTRATIONS

Case 1

A 48-year-old male patient presented with a 1 year history of right complete facial paralysis. The paralysis

Table 2. Preoperative and Postoperative Hearing Status of 20 Patients

<i>Hearing</i>	<i>Preoperative</i>	<i>Postoperative</i>
Normal	8*	8
Conductive hearing loss	1†	0
Sensorineural hearing loss	2	2
Dead ear	7	8
Data not available	2	—
Lost to follow-up	—	2

*One patient had a total deafness postoperatively.

†Patient regained normal hearing after a second-stage ossicular reconstruction.

Table 3. Various Surgical Approaches Used for Management of Intratemporal Vascular Tumors

<i>Approach</i>	<i>No.</i>
Middle fossa	5
Middle fossa-transmastoid	2
Transmastoid	2
Transmastoid extralabyrinthine	3
Translabyrinthine	5
Suboccipital	2
Transotic	1
Total	20

Table 4. Facial Nerve Function

Facial Nerve HB Grade*	Preoperative	Postoperative	
		Repaired	Intact
I	5	0	4
II	4	0	1
III	2	2	—
IV	4	2	—
V	2	4	—
VI	3	5 [†]	—
Lost to follow-up	—	2	—
Total	20	15	5

*HB: House-Brackmann.

[†]Three patients have follow-up less than 1 year because they were operated on recently.

was progressive in nature and was preceded with hemifacial spasm for 4 years. He had associated complaints of tinnitus for 6 months and had a single episode of vertigo. On examination, the tympanic membrane was normal and the facial nerve function was House-Brackmann (HB) grade VI. His audiovestibular evaluation, including auditory brainstem response (ABR) was normal. High-resolution CT scan with contrast medium showed a 5 mm mass at the region of tympanic segment and the second genu of the facial nerve with a modest contrast enhancement. MRI without contrast medium showed a diffuse mass of facial nerve in its intrapetrous course. The mass was isointense in T₁ images and hyperintense in T₂ images. MRI with gadolinium showed a discrete enhancement of the lesion in T₁ images. The transmastoid approach revealed vascular tumor of the facial nerve involving the intralabyrinthine portion, GG, tympanic portion, and the second genu. Intraoperative decision was taken to combine this approach with the middle fossa approach. A sural nerve graft was used to repair the facial nerve.

Histologic diagnosis was cavernous hemangioma. The postoperative course was uneventful. The patient is now in his third postoperative month and has his hearing preserved at the preoperative level.

Case 2

A 51-year-old male patient was referred to us with a 3-year history of progressive right facial nerve paralysis. He had no associated complaints of hearing loss, tinnitus, or vertigo. The clinical examination revealed a HB grade IV facial nerve paresis and audiovestibular investigations were within normal limits. High-resolution CT scan showed a mass involving the third portion of the facial nerve and the jugular bulb with irregular, indistinct bony margins (Fig. 1). MRI with gadolinium revealed a contrast enhancing mass in T₁-weighted images (Fig. 2). The patient underwent the transmastoid approach, which revealed a vascular tumor involving the facial nerve in its third portion with associated softening of the bone in relation to the tumor. A sural nerve graft was used for the repair of the facial nerve. Histologic diagnosis was cavernous hemangioma. Four months postoperatively, the patient maintained his preoperative hearing and it is still early to comment about the eventual functional outcome of the facial nerve.

Case 3

A 58-year-old patient presented with a 2-year history of right progressive facial nerve paralysis. The paralysis was preceded with hemifacial spasm. There were no associated complaints of hearing loss, tinnitus, or vertigo. Clinical examination showed a HB grade III facial nerve

Figure 1. Computed tomography scan of case 2 showing a vascular tumor involving the vertical portion of the facial nerve. Note the presence of diffuse demineralization and multiple bone radiolucencies.

Figure 2. Magnetic resonance image with gadolinium of case 2. T₁-weighted image showing a diffuse enhancing mass involving the facial nerve in its intratemporal course.

paresis. Audiovestibular investigation showed a mild right high tone SNHL with normal speech discrimination and ABR. High-resolution CT scan of the temporal bone revealed a soft tissue mass of the GG region (Fig. 3). The patient was recently operated on by a middle fossa approach. The tumor was situated at the GG with involvement of the intralabyrinthine portion of the facial nerve. Due to absence of plane of cleavage, the lesion was resected along with the involved portion of the facial nerve. The facial nerve was repaired using a sural nerve graft. Histologic diagnosis was hemangioma of arteriovenous type. The postoperative period was uneventful and the patient maintained his preoperative hearing level.

DISCUSSION

Intratemporal hemangiomas or vascular malformation in relation to the facial nerve were once thought to be rare, are now more often seen than the neuromas.^{9,10} Unfortunately, due to the variable clinical manifestation imitating the more common pathologic conditions, they are frequently misdiagnosed preoperatively. This delay in diagnosis often results in grave intratemporal consequences, such as hearing loss, facial nerve palsy,¹¹ or death from spontaneous hemorrhage into the epidural space.¹²

Figure 3. High-resolution computed tomography scan of case 3 showing vascular tumor at the geniculate ganglion.

Vascular lesions seem to arise from the vascular plexuses supplying or surrounding the facial nerve.^{13,15} The fact that these tumors compress the facial nerve from outside^{14,16} and in conjunction with the anatomic distinctness of the GG and geniculate capillary plexus, which is hypothesized to be the origin of most hemangiomas involving the facial nerve,¹⁵ perhaps speaks in favor of these tumors originating from outside rather than within the facial nerve.

Saleh et al,⁶ in a review of 41 cases of the vascular malformation of the IAC reported in the English literature, found most of these tumors initially presented with SNHL of retrochochlear type followed by facial nerve dysfunction and dizziness. The origin of these vascular lesion in the IAC has been postulated from the rich vascular plexus surrounding Scarpa's ganglion.¹⁴ This may explain the initial hearing loss of retrochochlear type rather than facial nerve dysfunction in these lesions.

In accordance with other authors,^{6,13,17} the tumors situated in the IAC were not distinguished preoperatively from an acoustic tumor. Although MRI appears sufficiently sensitive for detection of small intracanalicular tumors, distinction from an acoustic tumor is still difficult in individual cases.^{6,13} Furthermore, in a patient with radiographically revealed lesion of IAC along with the presence of facial nerve deficit associated with other clinical manifestations similar to acoustic tumor, one should consider the possible diagnosis of a vascular lesion.

Vascular lesions of the GG commonly present with facial nerve symptoms when of extremely small size.¹⁷ This may be attributed to the extraneural compression associated with direct invasion of the neural tissue.^{4,14} In the present study, facial nerve paresis was either preceded or accompanied with hemifacial spasm. Hemifacial spasm, which is not usually seen in patients with Bell's palsy, suggested the neoplastic involvement of the facial nerve.^{18,19} Moreover, the authors agree with Jackson et al¹¹ that progressive (lasting more than 4 weeks) facial weakness should be considered to have a tumor involving the facial nerve until proved otherwise.

On high-resolution CT vascular tumors when situated at the GG region typically show irregular, indistinct bone margins termed "honeycomb bone" and intratemporal bone spicules.¹⁰ In contrast, schwannomas of the facial nerve have a characteristic appearance of an well-defined, enhancing soft tissue mass, usually in the perigeniculate region with sharp bony erosion and enlargement of the fallopian canal.²⁰ The presence of diffuse, mottled demineralization with multiple honeycomb bone radiolucencies enabled us to diagnose the last three cases preoperatively.

Generally, the treatment aims at total removal of tumor with preservation of facial nerve functions and conservation of hearing. In the present series, hearing preservation or restoration could be achieved in the majority of cases that presented with preoperative serviceable hearing. Although preservation of facial nerve in

anatomic continuity was possible in five cases (all the tumors were located in the IAC), resection and repair of the facial nerve was required in 12 cases due to absence of clear plane of cleavage. Nine of 12 patients (75%) followed up for at least 1 year showed acceptable facial nerve function (HB grade IV or less).

CONCLUSION

Intratemporal vascular malformation involving the facial nerve are rare lesions. Due to the variable clinical manifestation imitating the more common pathologic conditions, they are frequently misdiagnosed preoperatively.

High-resolution CT scan, MRI with gadolinium, and a high index of suspicion are needed to diagnose these cases.

Early resection of these tumors enables better facial nerve outcome.

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