Intradural Facial Nerve Schwannoma: Diagnostic and Therapeutic Problems

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ABSTRACT

Objectives: To define characteristics of intradural facial schwannoma (FS) and discuss management strategies. **Method:** Retrospective review of clinical charts. **Results:** Six patients were diagnosed, based on surgical findings, as having intradural FS. None of the patients complained initially of facial weakness, but all showed severe disturbance of hearing function assessed on admission. FS was suspected in four of our six patients because of imaging findings or facial palsy caused by a small tumor. Total removal was performed in five cases followed by facial nerve reconstruction, and nearly total removal was performed in one case. Most patients recovered to Grade II or III palsy during the follow-up, and long-lasting palsy before surgery showed poor recovery. **Conclusion:** Imaging findings such as tumor extension to the middle fossa and clinical findings such as facial palsy caused by a small tumor strongly suggest a diagnosis of FS. Facial function can be restored satisfactorily for most patients after appropriate surgery.

KEYWORDS: Cerebellopontine angle, facial nerve, schwannoma, surgery

Tumors at the cerebellopontine angle (CPA) usually are vestibular schwannomas, meningiomas, and epidermoid tumors, but occasionally tumors with a different pathology can occur in this region, such as schwannomas originating from other cranial nerves, among which facial schwannoma (FS), which is often called facial neurinoma, occupies a special place. Its diagnosis and management are difficult, especially when it occurs intra-

durally, although the schwannoma may originate from any segment of the facial nerve. Sherman and colleagues¹ stated in a review of the literature that tumors limited to the CPA/internal auditory canal (IAC) constitute a unique subset of facial neuromas with characteristics that differ from those of facial neuromas in other locations and are clinically indistinguishable from acoustic neuromas. Thus surgeons may unexpectedly first find an FS during

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surgery of a CPA tumor that was thought preoperatively to be a vestibular schwannoma. Surgical results for vestibular schwannomas have recently improved, while preservation of not only facial but also hearing functions can be achieved in the case of small tumors. When dealing with FS, however, even facial function is at great risk. Surgeons are therefore faced with a difficult decision as to whether to remove the tumor completely in conjunction with reconstruction or to remove it partially and hope for recovery of residual functions.

For this article, we retrospectively reviewed cases with CPA tumors that we treated surgically in an attempt to define the characteristics of FSs arising at the CPA or IAC and for a discussion of management strategies.

PATIENTS AND METHODS

Between 1988 and 2004, the senior author (EK) surgically treated 166 CPA tumors. The clinical charts for these cases were reviewed retrospectively and data analyzed. Six patients were diagnosed from the surgical findings as having intradural FS located at the CPA and/or IAC. The criteria for the diagnosis were first, that the facial nerve, identified anatomically and by nerve stimulation if the patient had incomplete facial

palsy, extended to the tumor; second, that the eighth nerve could be separated easily from the tumor; third, that the tumor was identified histologically as a schwannoma.

Clinical pictures such as initial symptoms, facial and hearing function on admission, location of the tumor, surgical methods, and results were also analyzed. Facial function was evaluated with the House-Brackmann facial nerve grading system.²

RESULTS

Table 1 summarizes the characteristics of the six patients before surgery. There were four males and two females with a mean age of 52.3 years. It was noteworthy that none of the patients initially complained of facial weakness. For four patients the initial symptom was hearing disturbance. One of them (Case 2) presented with sudden deafness. The remaining two patients noticed altered taste sensation as the first symptom. However, facial palsy was noted on admission in three patients with House-Brackmann Grade III to IV. Two of these patients (Cases 2 and 6) had been admitted for the recent appearance of facial palsy. In the third patient (Case 3) complete facial palsy lasted for 2 years. The other three patients showed no facial motor weakness but two of them complained of diminished taste sensation. Hearing function assessed on admission by

Table 1 Preoperative Clinical Characteristics of the Six Patients with Facial Schwannoma

Case	Age	Sex	Side	Location	Initial Symptom	VII on Admission* (duration)	PTA on Admission
1	52	M	R	IAC-Gen	Hearing ↓	I	NR
2	43	M	L	IAC-Gen	Sudden deafness	III (2 mos)	60 dB
3	43	M	R	IAC	Hearing ↓	VI (2 yrs)	NR
4	54	F	L	CPA-IAC	Taste ↓	I, Taste ↓	60 dB
5	66	M	R	CPA-IAC	Taste ↓	I, Taste ↓	53 dB
6	56	F	R	CPA-IAC	Hearing ↓	III (3 mos)	75 dB

*Evaluated with House-Brackmann facial nerve grading system.

VII, facial nerve function in House-Brackmann Grade; PTA, pure-tone audiogram; M/F, male/female; R/L, right/left; IAC, internal auditory canal; Gen, geniculate ganglion; NR, no response; CPA, cerebellopontine angle.

means of pure-tone audiogram showed severe disturbance in all patients. Two patients were deaf and all the others had hearing loss of over 50 dB.

Location of the tumor was determined with high-resolution computed tomography (CT) and magnetic resonance imaging (MRI) with gadolinium (Gd) enhancement. The tumor was located from the IAC to the geniculate ganglion (Fig. 1) in two patients (Cases 1 and 2). In one patient (Case 3) the tumor location was confined entirely to the IAC (Fig. 2). The rest of the patients (Cases 4, 5, and 6) had tumors extending from the IAC to the CPA.

None of the patients had a family history of brain tumors. However, Case 5 had received a renal transplant 17 years previously and since then had continuously been taking immunosuppressants. In addition, 3 years previously she suffered from a pharyngeal cancer that was removed.

Selection of surgical approach, extent of tumor removal, method of reconstruction if performed, and facial function during the follow-up period of at least 2 years are listed in Table 2. In two patients (Cases 1 and 3), the translabyrinthine approach was selected as they had already lost hearing on the tumor side before surgery and FS was strongly suspected. The tumor was exposed with this approach and resected completely. The location of the healthy facial nerve could be identified as both proximal and distal to the tumor. In Case 1, the rerouting technique was used for re-

Figure 1 Gd-enhanced MRI of Case 1. No facial palsy was present on admission but the patient was deaf on the right side. MRI showed an inhomogeneously enhanced mass in the right IAC extending to the middle fossa (white arrow). FS was strongly suspected before surgery. Gd, gadolinium; MRI, magnetic resonance imaging; IAC, internal auditory canal; FS, facial schwannoma.

construction of the facial nerve. The distal end of the facial nerve was dissected as far as the vertical segment, released from the Fallopian canal, and anastomosed directly to the proximal end at the IAC without excessive nerve tension. A similar technique was tried for Case 3, but the defect was

Figure 2 (A) Gd-enhanced MRI and (B) bone-window CT of Case 3. (A) MRI showed a homogeneously enhanced tumor within the left IAC. (B) The IAC was markedly enlarged. FS was suspected because of a 2-year history of complete facial palsy and the intracanalicular location of the tumor. Gd, gadolinium; MRI, magnetic resonance imaging; CT, computed tomography; IAC, internal auditory canal; FS, facial schwannoma.

Table 2 Surgery and Facial Nerve Function

Case	Location	VII on Admission* (duration)	Surgical Approach	Extent of Removal and Method of Reconstruction	Final VII [†]
1	IAC-Gen	I	TL	Total, DS	II
2	IAC-Gen	III (2 mos)	TL	Total, Graft	III
3	IAC	VI (2 yrs)	RS + M	Total, Graft	VI
4	CPA-IAC	I, Taste ↓	RS	Nearly total	II
5	CPA-IAC	I, Taste ↓	RS	Total, DS	III
6	CPA-IAC	III (3 mos)	RS	Total, Graft	Too early [‡]

*Evaluated with House-Brackmann facial nerve grading system.

[†]Evaluated with House-Brackmann facial nerve grading system during follow-up of at least 2-years.

[‡]Grade IV palsy at 1-year follow-up.

VII, facial nerve function in House-Brackmann Grade; IAC, internal auditory canal; Gen, geniculate ganglion; TL, translabyrinthine approach; DS, direct suture; Graft, reconstruction with interposed sural nerve graft; RS, retrosigmoid approach; CPA, cerebello-pontine angle; M, mastoidectomy.

so long that an interposition graft taken from the sural nerve had to be used to reconstruct the facial nerve.

For the rest of the cases, the retrosigmoid approach was used to remove the tumor, an

approach routinely used by the senior author for the removal of CPA tumors. In Case 3, the tumor located solely within the IAC was exposed via the retrosigmoid approach and found to have originated from the facial nerve. The tumor was

Figure 3 (A,B) MRI and (C,D) intraoperative photos of Case 5. (A) Gd-enhanced MRI with fat suppression showed a multicystic tumor extending from the IAC to the right CPA. (B) T2-weighted image showed high and low cyst density. An old hemorrhage was detected during surgery. (C) After debulking of the tumor (T), the vestibulocochlear nerve (*) was easily separated from the tumor and the facial nerve (white arrow) was found to be the origin of the tumor. The facial nerve was dissected from the tumor as much as possible followed by total removal of the tumor. (D) Direct end-to-end suture could be performed at the CPA (white arrow) owing to the short segment of the defect and to the nerve redundancy caused by the large size of the tumor. Anatomical continuity of the vestibulocochlear nerve (*) was preserved.

removed completely but since it was not possible to find a healthy distal end of the facial nerve in the IAC, an additional mastoidectomy was performed to obtain the vertical segment of the facial nerve. A sural nerve graft was then anastomosed to the proximal stump at the CPA and to the distal stump at the mastoid. In Case 4, the tumor was removed nearly totally with preservation of most of the facial nerve. In Case 5 (Fig. 3) we tried to dissect the facial nerve from the tumor as much as possible and managed to remove the tumor completely while removing only a short segment of the nerve. Direct end-to-end suture at the CPA could be performed owing to the short segment of the defect and to the nerve redundancy caused by the large size of the tumor. In Case 6 (Fig. 4), the tumor was totally removed and a sural nerve graft was placed between the proximal end near the brain stem and the distal end at the IAC. A tiny tumor on the vagal nerve was found incidentally during the surgery.

Facial nerve function with the follow-up of at least 2 years is shown in Table 2. Although none of the patients could attain normal facial function, Grade II palsy was achieved in Cases 1 and 4 and Grade III palsy in Cases 2 and 5. Case 3 remained Grade VI after sural nerve grafting, possibly due to the 2-year history of complete facial palsy before surgery. Another case with sural nerve grafting has now been followed up for 1 year postoperatively. This patient cannot close one eye completely but has good recovery in the M. orbicularis oris (Grade IV). It is too early, however, for a final evaluation.

DISCUSSION

FS is a rare tumor and difficult to diagnose before surgery of CPA tumors. Sherman and associates¹ summarized 467 cases of FS from the literature including their own cases and found that 17.8%

Figure 4 T2-weighted image and intraoperative photos of Case 6. (A) The MRI showed a mainly high-intensity mass at the right CPA. (B) The tumor was exposed via the left retrosigmoid approach (T) and stimulation at the tumor surface produced strong EMG responses of the facial muscles. (C) The tumor was totally removed and a sural nerve graft (white arrow) was placed between the proximal end near the brain stem and the distal end at the IAC. (D) A tiny tumor was found incidentally on the vagal nerve.

originated at the CPA and 24.3% at the IAC segment. We identified six cases (3.6%) of FS among 166 CPA tumors consecutively operated on by a single surgeon. Intradural FS is thought to be clinically indistinguishable from vestibular schwannoma. Kubota and colleagues³ reviewed 28 cases of facial nerve schwannoma without facial palsy and found, very interestingly, that in 19 (67%) the tumor had arisen at the CPA. In our series, none of the patients had complained initially of facial weakness, which was indeed not apparent on admission in three patients, while two of them had noticed only altered taste sensation, so that their tumors may have originated from the intermediate nerve. Hearing function assessed by pure-tone audiogram on admission showed severe disturbance in all patients.

Although preoperative diagnosis of FS is usually difficult, we suspected it in four of our six patients. In Case 1, it was suggested by extension of the tumor from the IAC to the middle fossa while the patient had no facial palsy. In Case 2, FS was indicated by facial palsy even though the tumor was small and had extended to the middle fossa. In Case 3, suspicion of FS was based on a 2-year history of complete facial palsy while the tumor was limited to the IAC. In Case 6, possible diagnosis of FS was proposed because the patient developed facial palsy during preoperative follow-up of a small CPA tumor. Thus, imaging findings such as tumor extension to the middle fossa and clinical findings such as facial palsy caused by a small tumor are strong indications for a diagnosis of FS. Although FS could have been diagnosed in Cases 4 and 5 because of the altered taste sensation, our preoperative diagnosis was vestibular schwannoma.

Three of our patients already showed facial weakness on admission. One case had a long history of palsy while the other two had experienced recent episodes of facial palsy. For patients with facial palsy already present, radical removal of the tumor combined with reconstruction of the facial nerve should be the treatment of choice. It remains controversial, however, whether radical removal is advisable for patients without facial

palsy. Some authors advocate delaying surgery until the patient exhibits at least a House-Brackmann Grade III facial nerve paralysis.^{4,5}

If FS is suspected before surgery of a tumor at the CPA, treatment options can be discussed with the patient. These options comprise follow-up with serial imaging, total removal combined with nerve reconstruction, or subtotal removal, while radiosurgery is another possible therapeutic option⁶. It is not clear, however, whether the similarly good results by radiosurgery can be expected for FS as for vestibular schwannoma. If the patient's facial palsy is already more serious than Grade III, total removal and nerve reconstruction are recommended⁷. Liu and Fagan⁵ recommended delaying surgical removal for cases with mild or no facial dysfunction to maintain good facial function.

We followed up facial nerve function for at least 2 years and found that none of the patients could attain normal facial function. Sherman et al¹ reported that primary facial nerve preservation is often associated with an origin of tumor from nerves other than motor facial nerves, but preservation may be possible if the tumor is located eccentrically on the nerve. We could preserve facial nerve continuity in only one case managed with nearly total removal. Nerve suture with or without grafting is essential. Grade II palsy was achieved in one case treated with direct suture and one with nearly total removal. Repair with a sural nerve graft showed poorer results (Grades III and VI). Identical results of a final Grade III of facial expression have been reported for patients treated with nerve grafts.^{1,7} Improved results may be related to a better anastomosis. A long history of preoperative complete facial palsy could be the reason for the poor result of Case 3.

CONCLUSION

Intradural facial schwannoma is rare and difficult to diagnose before surgery. Imaging findings such as tumor extension to the middle fossa and clinical

findings such as facial palsy resulting from a small tumor are strong indications for a diagnosis of facial schwannoma. Facial nerve reconstruction is usually necessary in the case of tumor removal. Most patients treated with nerve reconstruction can be expected to recover to Grade II or III during the follow-up period.

REFERENCES

1. Sherman JD, Dagnew E, Pensak ML, van Loveren HR, Tew JM Jr. Facial nerve neuromas: report of 10 cases and review of the literature. *Neurosurgery* 2002;50:450–456
2. House JW, Brackmann DE. Facial nerve grading system. *Otolaryngol Head Neck Surg* 1985;93:146–147
3. Kubota Y, Kawamata T, Kubo O, Kasuya H, Muragaki Y, Hori T. Large facial nerve schwannomas without facial palsy: case reports and review of the literature. *Neurosurg Rev* 2005;28:234–238
4. Lipkin AF, Coker NJ, Jenkins HA, Alford BR. Intracranial and intratemporal facial neuroma. *Otolaryngol Head Neck Surg* 1987;96:71–79
5. Liu R, Fagan P. Facial nerve schwannoma: surgical excision versus conservative management. *Ann Otol Rhinol Laryngol* 2001;110:1025–1029
6. Hasegawa T, Kobayashi T, Kida Y, Tanaka T, Yoshida K, Osuka K. Two cases of facial neurinoma successfully treated with gamma knife radiosurgery [in Japanese]. *No Shinkei Geka* 1999;27:171–175
7. Chung JW, Ahn JH, Kim JH, Nam SU, Kim C-J, Lee K-S. Facial nerve schwannomas: different manifestations and outcomes. *Surg Neurol* 2004;62:245–252