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Ann Otol Rhinol Laryngol 2014 123: 647 originally published online 4 April 2014
DOI: 10.1177/0003489414528673

Annals of Otology, Rhinology & Laryngology
2014, Vol. 123(9) 647–653

DOI: 10.1177/0003489414528673
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Intraoperatively Diagnosed Cerebellopontine Angle Facial Nerve Schwannoma: How To Deal With It

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Abstract

Objective: This study aimed to report our experience in the management of patients with intraoperatively diagnosed intracranial facial nerve schwannomas (FNSs) and propose a decision-making strategy.

Methods: Twenty-three patients with FNS of the internal auditory canal and/or cerebellopontine angle operated on between 1992 and 2012 were identified.

Results: Preoperatively, all cases have been radiographically diagnosed as vestibular schwannomas. Operative procedures consisted of total tumor resection with grafting in 43.4% of patients, near-total resection leaving behind the tumor capsule overlying the facial nerve in 21.7%, total tumor resection with preservation of anatomic continuity of the facial nerve in 13%, and subtotal resection in 4.3%. Four patients (17.4%) underwent bony decompression with no tumor removal.

Conclusion: Management of FNS diagnosed at surgery represents a significant clinical challenge. We considered total tumor resection with grafting when patients presented with preoperative facial nerve palsy (≥ grade III). Both subtotal and near-total tumor removal can be performed in patients with preoperative good facial function and/or large tumors with brainstem compression. Patients with small tumors who were selected for hearing preservation surgery can be considered for bony decompression. Fascicle preservation surgery may be an option when a clear cleavage plane between the tumor and the facial nerve is found.

Keywords

facial schwannoma, facial nerve, cerebellopontine angle, internal auditory canal, facial nerve, management

Introduction

Facial nerve schwannomas (FNSs) are slow-growing rare benign tumors that can arise along any segment of the facial nerve from the cerebellopontine angle to the peripheral branches in the face and usually involve multiple nerve segments.[1-3] The most common locations have been reported to be the geniculate ganglion (68%), the labyrinthine (52%), and the tympanic (43%) segments.[4] Facial nerve schwannomas limited to the internal auditory canal (IAC) and/or the cerebellopontine angle (CPA) are less common, accounting for about 2% of all FNSs.[5]

Posterior cranial fossa FNSs usually manifest with sensorineural hearing loss, tinnitus, and vertigo.[5-7] Thus, the clinical presentation of these lesions mimics that of the more common vestibular schwannoma (VS). Moreover, in such a location, they frequently present without facial nerve dysfunction, often leading to establish the true diagnosis only during surgery. Once a FNS has been intraoperatively diagnosed, its management is particularly demanding especially in the presence of normal facial nerve function.

Until 2 decades ago, surgical removal with nerve grafting was considered the treatment of choice for FNS regardless of facial nerve function.[8] The profound cosmetic defect caused by facial nerve palsy could have a devastating psychological and clinical effect on the patient's life, questioning the relevance of surgical tumor removal in patients with no or minimal preoperative facial nerve dysfunction. Nowadays, gross tumor resection with facial nerve grafting

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is usually reserved for patients with poor preoperative facial nerve function or when serious intracranial complications become imminent.[8,9] In fact, the almost inevitable sacrifice of the facial nerve during surgery requires nerve reconstruction with subsequent postoperative paralysis, which is followed by some degree of recovery that will unlikely be better than House-Brackmann (HB) grade III. In recent years, several facial nerve preservation surgical procedures that include partial removal (tumor debulking), decompression, and complete tumor resection with facial nerve preservation (fascicle preservation surgery) have been developed to treat FNSs in the presence of good facial function.[8]

The aim of the present study is to report our strategy in the management of intraoperatively diagnosed intracranial FNSs based on our experience in treating 23 of these patients.

Methods

A retrospective review was performed of all patients with the diagnosis of VS treated by the senior authors (M.S. and J.M.) over a period of 20 years, from July 1991 to January 2012. Of 2292 IAC/CPA tumors, 23 (1%) were FNSs and the subjects of this study. A tumor was diagnosed as FNS if the facial nerve was clearly identified as the origin of the tumor at the time of operation. Only purely IAC and/or CPA FNSs were included in this study. Lesions that were found to extend to the geniculate ganglion were excluded from the study. The charts of the patients, including surgical records, neuroimaging studies, histological records, and follow-up records were reviewed retrospectively. Tumor size was evaluated by measuring the largest diameter of the extrameatal component alone on gadolinium-enhanced magnetic resonance imaging (MRI).[10] A tumor of 0 mm refers to a purely intracanalicular lesion. The tumor resection is defined as total when resection is complete, and as near-total when the tumor capsule or tiny remnants overlying the facial nerve are left behind. Subtotal removal refers to a minor residue that is left because the tumor was too adherent to the facial nerve and there was no distinguishable cleavage plane that could be followed, providing a harmless dissection without damaging the nerve. Preoperative and postoperative facial nerve function were assessed according to the HB grading system.[11] Hearing results were evaluated according to the guidelines of the American Academy of Otolaryngology–Head and Neck Surgery.[12] All patients were followed up at the outpatient clinic for at least 1 year after surgery.

Results

Patient's Characteristics and Clinical Features

A total of 23 patients with IAC/CPA FNS were identified and analyzed for the present study. They consisted of 8 males (34.8%) and 15 females (65.2%), ranging in age from 13 to 76 years (mean, 45.4 years). There was no predominance regarding the side of the lesion. Hearing loss (65.2%), tinnitus (43.5%), and vertigo (39.1%) were the most common presenting symptoms. Facial weakness was encountered only in 6 patients (26%). Among these, the facial paralysis was gradually progressive in 3 cases, and it was of sudden onset mimicking a Bell's palsy in the remaining patients. The delay between the first symptom and diagnosis ranged from 3 months to 12 years (mean, 36.2 months). Relevant information on the patients is provided in Table 1. The follow-up (consisting of clinical evaluation, hearing tests, and serial MRI scans) of the series ranged from 12 to 84 months (mean, 18 months). Follow-up was 100% complete.

Imaging Features

Preoperatively, all cases had been radiographically misinterpreted as VSs. Eight of the 23 patients (34.8%) had a tumor located only in the CPA. Two patients (8.7%) had a tumor limited to the IAC with no measurable extension into the CPA. Thirteen patients (56.5%) had a FNS in the IAC extending into the CPA. Of the 15 cases with IAC involvement, the tumor extended laterally into the fundus in 4 cases, whereas the latter was free in the other 11 cases. The tumors ranged in size from 0 mm (purely intracanalicular lesion) to 2.5 cm with a mean (SD) of 1.0 (0.78) cm.

Treatment and Outcomes

Two patients with only a slight dizziness at diagnosis and a purely intracanalicular tumor were initially managed conservatively with clinical and radiological observation. Both required subsequent surgery 4 and 6 years, respectively, after the diagnosis was made. One of them showed evidence of tumor growth on MRI, whereas the other one experienced a slowly progressive facial paralysis with evidence of tumor growth on repeated imaging. The remaining 21 patients underwent surgery as the first-line treatment. Ultimately, all patients in this study underwent surgery. The translabyrinthine approach was used for patients with large tumors or nonserviceable hearing (13 patients). Hearing preservation was attempted using the retrosigmoid approach in patients with smaller tumors where hearing was good or serviceable (10 patients).

Nineteen of the 23 patients had surgical excision, whereas the other 4 underwent bony decompression with no tumor removal. Among those who underwent surgical excision, anatomic integrity of the facial nerve could be preserved in 9 patients (39.1%). Three of them had total tumor removal with preservation of the nerve fibers (fascicle preservation surgery), 5 had near-total resection leaving behind the tumor capsule overlying the facial nerve fibers, and 1 had subtotal tumor removal. Ten patients who underwent

Table 1. Summary of Patients.

Case	Age/ Sex	Symptoms	Tumor Location	Size, cm	Procedure	Tumor Removal	FN Management	Preop. HB Grade	Last Examination HB Grade	Preop. Hearing Class AAO-HNS	Postop. Hearing Class AAO-HNS
1	43/F	FP, HL	CPA	1.5	RS	Total	VII-XII	III	V	B	DE
2	30/F	HL	IAC-CPA	0.5	RS	None	Decompression	I	I	B	B
3	45/F	V	IAC-CPA	0.3	RS	None	Decompression	I	I	A	A
4	44/F	V, T	IAC-CPA	0.5	RS	Total	FPS	I	II	A	DE
5	33/F	HL, T	IAC-CPA	0.7	RS	Total	FPS	I	I	B	DE
6	49/F	V, T	IAC-CPA	1.0	RS	None	Decompression	I	I	A	A
7	13/F	FP, HL	CPA	1.5	RS	Total	VII-XII	V	III	B	DE
8	59/M	HL, V	IAC-CPA	0.8	RS	Near-total	None	I	I	B	DE
9	46/M		IAC-CPA	0.7	RS	Total	Sural graft	I	III	A	DE
10	45/F	HL	IAC-CPA	0.5	RS	None	Decompression	I	I	B	B
11	58/F	FP, HL	IAC-CPA	2.0	TL	Total	GAN graft	V	III	D	DE
12	30M	FP, HL, T	IAC	0	TL	Total	GAN graft	I	III	D	DE
13	76/M	FP, HL	IAC-CPA	2.0	TL	Total	VII-XII	IV	III	D	DE
14	55/F	V	CPA	2.4	TL	Total	FPS	I	III	A	DE
15	54/M	HL, V	IAC-CPA	0.5	TL	Near-total	None	I	I	D	DE
16	52/F	FP, V, T	CPA	2.5	TL	Near-total	None	I	I	A	DE
17	59/M	FP, HL	CPA	0.8	TL	Near-total	None	I	I	C	DE
18	53/F	HL	CPA	2.5	TL	Total	Sural graft	I	III	DE	DE
19	32/F	HL, T	IAC	0	TL	Near-total	None	I	II	C	DE
20	24/M	HL	IAC-CPA	0.5	TL	Total	Sural graft	I	IV	D	DE
21	46/M	V, T	CPA	2.0	TL	Total	Sural graft	I	III	A	DE
22	47/F	HL, V, T	CPA	1.2	TL	Total	Sural graft	I	III	C	DE
23	53/F	Facial twitching, T	IAC-CPA	1.5	TL	Subtotal	None	I	I	C	DE

Abbreviations: AAO-HNS, American Academy of Otolaryngology–Head and Neck Surgery; CPA, cerebellopontine angle; DE, dead ear; FN, facial nerve; FP, facial nerve palsy; FPS, fascicle preservation surgery; GAN, great auricular nerve; HB, House-Brackmann; HL, hearing loss; IAC, internal auditory canal; RS, retrosigmoid approach; T, tinnitus; TL, translabyrinthine approach; V, vertigo.

complete tumor resection had the facial nerve sacrificed: the facial nerve was reconstructed either with grafting (7 cases) or hypoglossal-facial anastomosis (3 cases). The latter technique was used when the proximal stump of the severed facial nerve was too close to the brainstem. Facial nerve results are detailed in Table 1. Facial nerve results presented according to tumor treatment are displayed in Table 2. Hearing was preserved at the preoperative level in 4 patients (cases 2, 3, 6, and 10), whereas hearing was lost in the other 6 cases.

Of the 10 patients without total removal, only 1 patient experienced growth at 24 months of follow-up. To date, no recurrence has been observed at neuroradiologic controls in the 13 cases of total tumor resection.

Table 2. Facial Nerve Outcomes According to Treatment.

Treatment	No. of Cases	Facial Nerve Function HB Grade					
		I	II	III	IV	V	VI
TRWR (cable grafting)	7[a]	—	—	6	I	—	—
TRWR (VII-XII)	3[b]	—	—	2	—	I	—
FPS	3	I	I	I	—	—	—
Near-total removal	5	4	I	—	—	—	—
Subtotal removal	I	I	—	—	—	—	—
Decompression	4	4	—	—	—	—	—

Abbreviations: FPS, fascicle preservation surgery; HB, House-Brackmann; TRWR, total tumor removal with nerve repair.
[a]One patient had preoperative HB grade V and reached grade III 1 year after surgery.
[b]Preoperatively, 1 patient had HB grade III, 1 had HB grade IV, and 1 had HB grade V.

Discussion

Facial nerve schwannomas isolated to the IAC and CPA are rare, accounting for approximately 1% to 3% of all CPA

Table 3. Review of the Largest Series of Cerebellopontine Angle Facial Nerve Schwannomas.

Study	No. of Cases	Tumor Location	Preop. FN Palsy, No. (%)	Treatment	Surgical Approaches	FN Reconstruction	Postop. HB
McMenomey et al[6]	12	CPA, 6 IAC-CPA, 5 IAC, 1	0	TRWR, 7 FPS, 5	TL, 9 TL-MCF, 1 RS, 2	Primary anastomosis, 2 VII-XII, 5	II, 2 III, 6 IV, 1 V, 3
Sherman et al[14]	6	IAC, 1 CPA, 3 IAC-CPA, 2	2 (33.3)	TRWR, 2 FPS, 2 Decompression, 1	TL, 2 RS, 2 MCF, 1	Cable grafting, 2	I, 3 II, 1 III, 1 V, 1
McMonagle et al[18]	16	IAC, 2 CPA, 6 IAC-CPA, 8	5 (31.2)	TRWR, 3 FPS, 1 SR, 4 Decompression, 1 Open and close, 5 Observation, 2	TL, 9 RS, 5	Primary anastomosis, 2 Cable grafting, 1 VII-XII, 1	I, 9 II, 2 III, 5
Lee and Kim[26]	5	IAC, 3 IAC-CPA, 2	1 (20)	FPS, 4 Open and close, 1	TL, 3 MCF, 2	—	I, 2 II, 3
Mowry et al[5]	16	IAC, 9 CPA, 4 IAC-CPA, 2	1 (6.2)	TRWR, 2 FPS, 2 SR, 10 Decompression, 2	TL, 8 MCF, 8	Cable grafting, 2	I, 9 II, 4 III, 1 IV, 1
Nadeau and Sataloff[13]	6	IAC-CPA, 6	0	TRWR, 3 FPS, 3	TL, 6	Primary anastomosis, 1 Cable grafting, 2	II, 2 III, 2 IV, 1 V, 1
Present study	23	IAC, 2 CPA, 8 IAC-CPA, 13	6 (26)	TRWR, 10 FPS, 3 SR, 1 NTR, 5 Decompression, 4	TL, 13 RS, 10	Cable grafting, 7 VII-XII, 3	I, 10 II, 2 III, 9 IV, 1 V, 1

Abbreviations: CPA, cerebellopontine angle; FN, facial nerve; FPS, fascicle preservation surgery; HB, House-Brackmann scale; IAC, internal auditory canal; MCF, middle cranial fossa approach; NTR, near-total removal; RS, retrosigmoid approach; SR, subtotal removal; TL, translabyrinthine approach; TRWR, total tumor removal with facial nerve reconstruction.

tumors.[5-7,13] We reviewed our experience with IAC/CPA FNSs from 1991 to 2012. Out of a total of 2292 IAC/CPA tumors treated during this period, only 23 (1%) were FNSs. Very few series have been reported that included more than 5 cases. A summary of treatment and outcome in studies reporting 5 or more patients with IAC/CPA FNSs is reported in Table 3.

The rarity of IAC/CPA FNSs and their clinical and radiological presentation make these lesions often misdiagnosed as VS. Clinically, the symptoms caused by intracanalicular and CPA FNSs replicate those of the more common VSs, with hearing loss being the most common presenting symptom along with tinnitus and vertigo.[5,6,13-16] Facial dysfunction is a less frequent presenting symptom in FNSs isolated to the IAC and CPA.[5,14,15] This can be explained by the fact that the tumor must reach a substantial size to cause significant nerve compression with subsequent dysfunction.[13] However, all patients with facial dysfunction and an IAC/CPA mass should be suspected to have a FNS, as only 1% to 4.7% of cases with VS have this symptom.[17] In our series, only 26% of the patients presented initially with facial weakness, whereas 65.2% had some degree of hearing loss. A similar finding was reported by other authors.[14,16]

At present, MRI cannot differentiate posterior cranial fossa FNS from VS if no extension into the labyrinthine segment of the facial nerve is present.[15,18] In such cases, MRI will show an enhancement along the labyrinthine segment of the facial nerve, producing the so-called "labyrinthine tail sign." Patients without preoperative facial nerve symptoms and absence of the labyrinthine tail are usually misdiagnosed preoperatively. Fagan et al[19] have described another radiological feature to distinguish FNS from VS. They noted in all 4 of their cases of FNSs confined to the IAC and CPA that the bulk of the tumor occupied a position eccentric to the axis of the IAC. However, this finding was not found either in our series or in those of others.[14,20]

Surgical resection with facial nerve reconstruction is usually the standard management for patients with facial

nerve function of HB grade III or worse. In fact, surgical tumor removal requires facial nerve repair in the majority of cases and, until now, no reconstruction method has been able to guarantee facial function recovery better than HB grade III.[3,5,8,14,18,21,22] This was clearly evident in 6 of our patients with normal preoperative facial function in whom we performed surgical excision and grafting. At final follow-up, 5 patients reached HB grade III, and 1 had HB grade IV. A number of facial nerve preservation surgical procedures that include partial tumor removal, bony decompression, and fascicle preservation surgery have been developed to maximize facial nerve function as long as possible.[8,18] Partial resection has the advantage of anatomic preservation of the nerve but has the risk of postoperative facial palsy.[5,6,23] The main drawback of partial resection is the difficulty of establishing where the removal should be stopped to preserve facial function. A recent literature review of 39 patients who underwent debulking for FNS showed that 26 (66.6%) patients achieved HB grade I to II facial nerve function, 10 patients (25.6%) had HB grade III, and only 3 patients (7.7%) had HB grade V to VI.[5] In our series, 1 patient had a subtotal resection and 5 others had a near-total removal leaving behind the tumor capsule overlying the nerve fibers. Five of these patients (83.3%) maintained their preoperative normal facial function and 1 experienced HB grade II.

Surgical resection with nerve preservation, also termed *fascicle preservation surgery*, aims to preserve the main trunk of the facial nerve and was first reported by Pulec[24] in 1972 for treatment of small intratemporal FNSs. McMenomey et al,[6] in 1994, and Sataloff et al,[25] in 1995, used this technique for selected cases of FNSs involving the CPA. Since then, other authors have performed fascicle preservation surgery for resection of selected FNSs in patients with good preoperative facial nerve function.[13,16,23] This technique is based on the finding that at the beginning of its growth, FNS grows eccentrically from the nerve involving only a small portion of nerve fibers. In larger tumors, the nerve fibers are often found within the tumor mass. In only 3 of our patients, we were able to find a clear plane between the tumor and the facial nerve and completely separate the tumor from the perineurium of the nerve. However, only 1 of these patients maintained normal facial function, whereas 1 had HB grade II and 1 experienced HB grade III. These results are consistent with those reported in the literature.[6,13,26] This raises the question of whether using facial nerve preservation procedures in patients with normal nerve function is a viable option or not.

Facial nerve decompression was first proposed by Angeli and Brackmann[20] in 1997 to reduce interfascicular pressure produced by a growing tumor and stabilize facial function. This technique allows patients to maintain their preoperative facial nerve function. Angeli and Brackmann[20] demonstrated excellent outcomes in a series of 4 unsuspected IAC/CPA FNSs undergoing decompression. Three patients maintained HB grade I, and the remaining patient with preoperative grade V achieved HB grade II. Of these 4 tumors, 3 cases had no tumor growth after 24, 36, and 60 months, whereas 1 increased 6 mm in almost 5 years. We performed decompression of the IAC through a retrosigmoid approach in 4 patients with normal preoperative facial function. Facial function was preserved in all of them.

Establishing a correct preoperative diagnosis of FNS would be fundamental for treatment planning and patient counseling. However, distinguishing an intracranial FNS from VS preoperatively may be extremely difficult even after a thorough neuroradiological workup. The correct diagnosis is often made during surgery, which poses a dilemma to the surgeon: remove the tumor and cause an unexpected facial palsy or abort the procedure.

Our experience with IAC/CPA FNSs has led us to propose a treatment algorithm for FNSs diagnosed at surgery (Figure 1).

Total tumor resection with grafting was considered when patients presented with preoperative facial nerve palsy (≥ grade III). Both subtotal and near-total tumor removal can be performed with excellent outcomes in patients with preoperative good facial nerve function (HB grade I-II) and large tumors with brainstem compression. Patients with small tumors selected for hearing preservation surgery should be considered for bony decompression. Fascicle preservation surgery may be an especially attractive option when a clear cleavage plane between the tumor and the facial nerve is found. It is important to note that both the decompression and partial tumor resection procedures leave the tumor on the facial nerve with the need for serial postoperative MRI to assess for tumor regrowth.

In the unlikely circumstance where the diagnosis of IAC/CPA FNS is ascertained preoperatively, the main management strategies described in the literature include (1) observation, (2) radiation, and (3) surgery. Since FNSs are generally slow-growing tumors and facial function is often normal and remains stable for long periods, observation with repeated imaging is a reasonable option as an initial management strategy.[9,17,18,20,27-29] Surgery is indicated when facial function deteriorates to HB grade III or in the presence of intracranial complications such as hydrocephalus and brainstem compression. Recently, radiotherapy has been advocated as a primary therapeutic option in patients with FNS.[30-35] The efficiency of radiotherapy in long-term tumor control is still unknown, and further studies with longer follow-up are necessary.[9] The advantages of radiotherapy include avoidance of surgery and possible arrest of tumor growth. The disadvantages include persistence of the tumor, risk of failure to control the growth, and malignant degeneration.[8-36] Therefore, although radiotherapy is considered an attractive conservative treatment modality in FNS, the above-mentioned suspicions should be considered

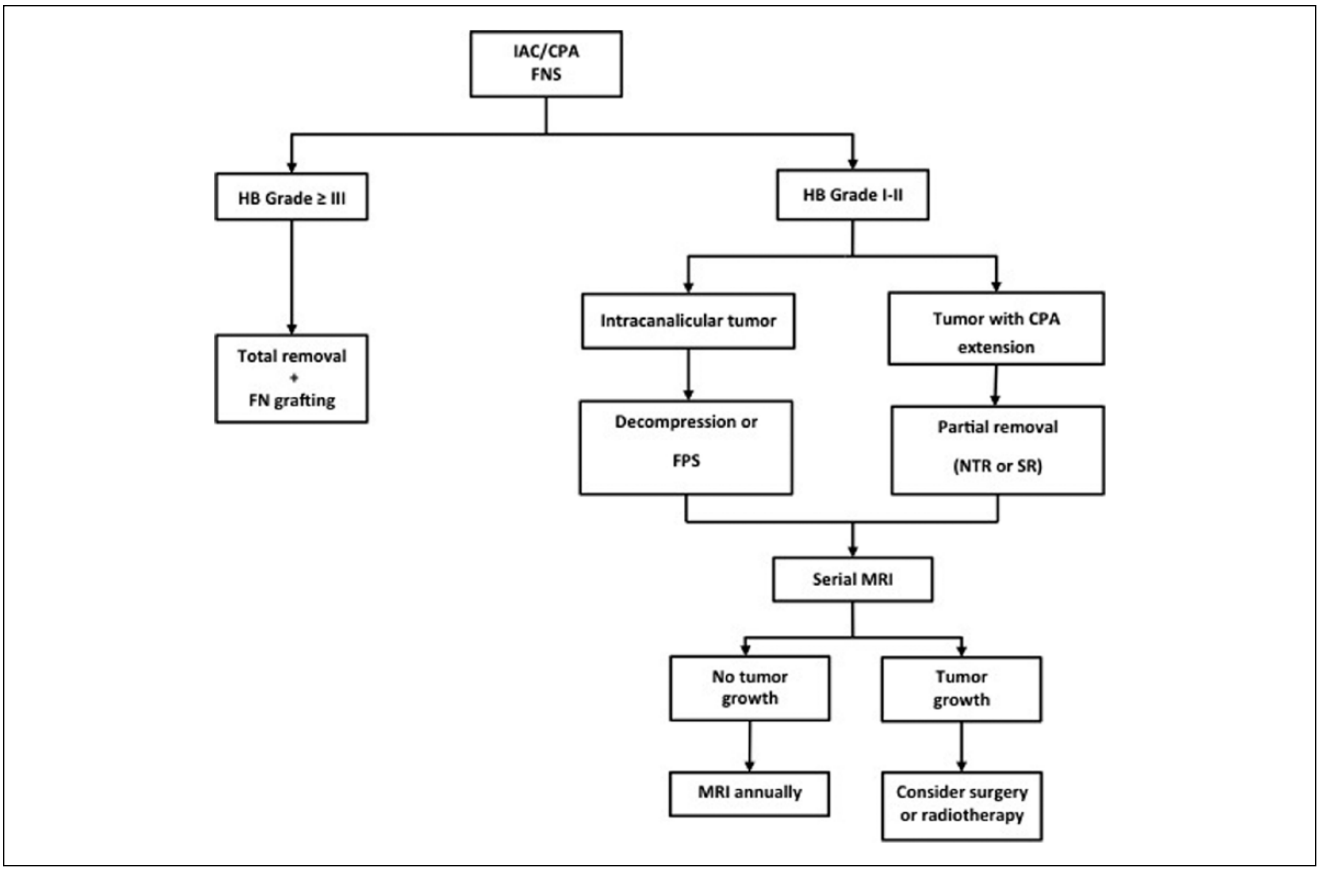

Figure 1. Algorithm for the management of intraoperatively diagnosed intracranial facial nerve schwannomas. CPA, cerebellopontine angle; FN, facial nerve; FNS, fascicle nerve schwannoma; FPS, fascicle preservation surgery; IAC, internal auditory canal; MRI, magnetic resonance imaging; NTR, near-total removal; SR, subtotal removal.

in the counseling and the decision-making process for the treatment of such patients. At present, we do not consider radiotherapy as an adequate primary treatment policy for these tumors. We reserve it as an alternative to surgery in (1) selected patients in whom there are serious contraindications to surgery (ie, critical general medical conditions, elderly patients); (2) patients with tumor regrowth after subtotal resection or decompression; and (3) patients who refuse surgery.

Conclusion

Internal auditory canal/CPA FNSs pose a major diagnostic difficulty, usually mistaken for VSs. Apart from the few cases in which tumor extension to the labyrinthine segment could establish the diagnosis, most cases are actually diagnosed intraoperatively. In the presence of an IAC/CPA lesion, we recommend considering the possibility of a facial nerve tumor.

The management of FNSs has evolved over the years from radical excision and grafting toward a more conservative treatment. We consider total tumor removal as the primary treatment to undertake in patients with a facial weakness worse than a grade III of HB. In patients with no or mild facial nerve dysfunction, we recommend conservative surgical management either through bony decompression or partial tumor removal.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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