Management of Hypoglossal Schwannomas: Single Institutional Experience of 14 Cases

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Abstract

Background Hypoglossal schwannomas are rare intracranial neoplasms. Microsurgical resection with the goal of cure is the aim of management but is associated with a high rate of postoperative morbidity.

Objective The objective of the study was to outline the clinical presentation, radiologic characteristics, surgical techniques, postoperative morbidity, and long-term follow-up results for hypoglossal schwannomas.

Methods Patients treated for hypoglossal schwannoma at the Department of Neurosurgery of a tertiary-level referral institution from January 2001 until December 2010 were analyzed retrospectively using hospital records.

Results There were 14 patients who were treated in the study period. Tongue atrophy and swallowing difficulties were the most common presenting symptoms. Surgery done in 12 patients using a variety of approaches (retromastoid retrosigmoid suboccipital in 9, midline suboccipital in 2, and far lateral in 1). Five patients having small residual tumors received gamma knife (GK) subsequently. Two patients received primary GK stereotactic radiosurgery. Three patients had permanent morbidity in the form of cranial nerve paresis. Immediate postoperative complications like cerebrospinal fluid leak and pneumonia were present in three patients.

Conclusion Complete microsurgical resection is often associated with a high rate of morbidity. Subtotal and near-total resection followed by stereotactic radiosurgery or observation now offers an alternative approach.

Keywords

- ▶ hypoglossal canal
- ▶ schwannoma
- ▶ skull base tumors
- ▶ gamma knife
- ▶ twelfth nerve

Introduction

Hypoglossal schwannomas are extremely rare cranial base neoplasms arising from Schwann cells of the 12th cranial nerve (CN)¹ accounting for only 5% of all nonacoustic intracranial schwannomas.² Hypoglossal schwannomas (neurinomas) were reported for the first time by De Martel in 1933.¹ These tumors are most often seen in middle-age women and can be completely intracranial, intracranial/extracranial, or completely extracranial.³ Symptoms differ, depending on the location of the tumor. Tongue wasting is present for most patients at the

time of presentation.^{4–6} Suboccipital headache, exacerbated by neck movement, are early symptoms of intracranial tumors.⁷ Patients with extracranial tumors may present with a mass in the upper neck or parapharyngeal area, in addition to wasting of the tongue.³ Larger tumors also present with cerebellar and brainstem signs. These symptoms seem to be correlated closely with tumor bulk of the intracranial portion. Dumbbell-shaped tumors with extension to the jugular foramen may also exhibit jugular foramen syndrome.

These tumors are benign slow-growing tumors; thus complete resection affords cure. As with all CN schwannomas,

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Table 1 Demographic profile and clinical presentation of hypoglossal schwannoma ($n = 14$)

Patient no.	Age, y/sex	Side	Clinical signs									
			Cranial nerve palsy							Pyramidal signs	Cerebellar signs	Impaired vision
			III/IV/VI	V	VII	VIII	IX/X	XI	XII			
1	24/M	B/L		+		+	+		+	+	+	+
2	30/F	R						+	+			
3	63/F	L							+		+	
4	61/F	R		+	+	+	+		+	+	+	
5	23/M	R	+				+		+		+	
6	35/M	L			+		+	+	+	+	+	
7	16/F	R					+	+	+		+	
8	42/M	R					+		+			
9	35/F	L			+	+	+		+			
10	43/M	R					+		+	+	+	
11	34/M	L		+	+	+	+	+	+		+	
12	15/M	R							+			
13	53/F	R							+			
14	51/M	R					+		+			

operative morbidity and mortality is caused by brainstem compression and associated CN palsies.^{4–6} No racial or sexual predilection has been described in these schwannomas. The surgical approach that is most appropriate for these tumors remains controversial. The literature about hypoglossal schwannomas is sparse and mostly case reports. To date, ~160 cases of hypoglossal schwannomas have been reported in the English literature.⁸ We report single-institution experience of management of 14 cases of hypoglossal schwannomas. The present series is one of the largest series of hypoglossal schwannomas in the existing English medical literature.

Subjects and Methods

Of > 1,000 patients with cerebellopontine tumors including schwannoma, meningioma, and other tumors referred to our center between January 2001 and December 2010, only 14 were treated for hypoglossal schwannoma in the Department of Neurosurgery of a tertiary-level referral institution.

Case files, radiologic scans, and outpatient records were retrospectively analyzed and demographic data (age, gender, and clinical presentation), radiologic findings (size and extent of tumor and associated hydrocephalus), whether surgical or radiosurgical management (if surgical, extent of surgical resection and postoperative complications) were recorded. Follow-up data included any permanent neurologic deficits, recurrent/residual tumor, and any adjuvant therapy or redo surgery. Surgical approaches were chosen depending on the primary location and extent of the tumor. Large tumors were operated on, and small tumors (< 3 cm in size) were usually given gamma knife (GF) treatment.

Results

A total of 14 patients were treated during the study period; eight (57.1%) were male and six (42.8%) were female. Mean age of the patients was 37.5 years with a range of 15 to 63 years. The right side (9 patients [64.2%]) was involved more commonly than the left (4 patients [28.5%]); one patient had a bilateral lesion. Two patients (14.2%) had type 2 neurofibromatosis as seen in other series also.⁹

The duration of symptoms ranged from 1 to 6 years with a mean of 2.3 years. The most common presentation was CN deficits, with the most common hypoglossal nerve involvement in the form of tongue fasciculation and atrophy (12 patients [85.7%]). Ten patients (71.4%) had involvement of CN IX and X. Four patients (28.5%) had preoperative facial nerve paresis. Patients with larger tumors presented with cerebellar (eight patients [57.1%]) and pyramidal tract signs (four patients [7.1%]) (► **Table 1**).

Twelve patients underwent definitive surgery, and two patients (with tumors < 3 cm in size) were given primary GF treatment. Retromastoid retrosigmoid suboccipital craniotomy with opening of foramen magnum for intradural tumors with large posterior fossa component was performed in nine cases (75%). Midline suboccipital craniotomy was used in two cases (16.6%). For large tumors with caudal extension through the foramen magnum, a far lateral approach was used in one case (8.3%) for transdural and extradural dumbbell-shaped tumors. There was no case of extracranial cervical extension of tumor in our series. In five patients (41.6%), complete tumor excision was performed; near-total excision was achieved in six patients (50%) and only subtotal decompression in one patient (8.3%) (► **Fig. 1**).

Fig. 1 (A, B) Preoperative T1-weighted contrast magnetic resonance images showing a heterogeneously hyperintense tumor in the hypoglossal canal with large posterior fossa component. (C, D) Postoperative images showing small residual tumor in the hypoglossal canal for which adjuvant gamma knife therapy was given.

In five patients (41.6%), complete tumor excision was performed; near-total excision was achieved in six patients (50%) because part of the tumor was adhered to the brainstem. Only subtotal decompression was achieved in one patient (8.3%) (→ **Table 2**).

Eight patients had an uneventful postoperative recovery. Three patients (25%) had a prolonged postoperative course with chest infection in two (16.6%). In our series, one patient developed postoperative cerebrospinal fluid leak from the surgical site that could have been avoided by dural augmentation and the use of tissue glue, although there was no hydrocephalus on preoperative scans.

Transient deficits of IX and X CNs were present in 8 patients (66.6%) with two patients (16.6%) requiring tracheostomy. No improvements in preoperative CN deficits were noted in any cases. Permanent morbidity, in the form of facial paresis, was seen in three patients, maybe because of their large size and associated cystic component. These could have been avoided by doing near-total or subtotal resection of tumor and subsequent treatment by radiosurgery. In the presence of intraoperative detection of facial nerve avulsion, primary sural nerve interposition grafting can also be tried.⁸

There was no mortality in the series.

All patients who underwent surgery had histologically verified benign schwannomas. The follow-up period ranged from 3 to 59 months, with a mean of 32 months. All patients had follow-up imaging that revealed residual tumor in five cases (41.6%), for which adjuvant GF therapy was given.

GF therapy was performed using a Leksell Gamma Knife (Elekta Instruments, Atlanta, GA, USA), with a tumor margin dose of 12 to 13 Gy, to patients at the Gamma Knife Center in the Department of Neurosurgery. Primary GK treatment was given to 2 patients (28.5%), and five patients (71.4%) received GK in the postoperative period for small residual tumors. All seven patients who had undergone GK had good tumor control with five patients (71.4%) having their tumor size unchanged and two (28.5%) having reduced tumor size.

Discussion

The surgical resection of hypoglossal schwannomas was associated with high mortality and morbidity in the literature before the 1970s. Over 50% of the patients died of respiratory difficulty within 4 weeks after surgery.^{3,7,10} Because of

Table 2 Treatment summary of hypoglossal schwannoma in our series ($n = 14$)

Patient no.	Type of tumor as per classification	Extent of tumor	Approach	Extent of resection	Post operative outcome	Auxiliary procedure
1	Type A	Intradural with caudal extension through the foramen magnum	Midline suboccipital craniotomy	Total	CSF leak, facial palsy	None
2	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Total	Uneventful	None
3	Type A	Intradural with caudal extension through the foramen magnum	Midline suboccipital craniotomy	Total	Uneventful	None
4	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Near total	Chest infection	Tracheostomy
5	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Near total	Facial palsy	Secondary radiosurgery
6	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Subtotal decompression	Uneventful	Secondary Radiosurgery
7	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Near total	Uneventful	Secondary radiosurgery
8	Type B	Transdural and extradural dumbbell-shaped tumor	Far lateral	Total	Uneventful	None
9	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Near total	Uneventful	None
10	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Total	Chest infection, facial palsy	Tracheostomy
11	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Near total	Uneventful	Secondary radiosurgery
12	Type A	Intradural with large posterior fossa component	Retromastoid/sigmoid suboccipital craniotomy	Near total	Uneventful	Secondary radiosurgery
13	Type A	Intradural tumor	Primary radiosurgery	–	Uneventful	None
14	Type A	Intradural tumor	Primary radiosurgery	–	Uneventful	None

Abbreviation: CSF, cerebrospinal fluid.

frequent postoperative respiratory difficulties, a mandatory prophylactic tracheostomy had been recommended before 1970.^{3,7} With the development of micro-neurosurgical techniques, use of adequate monitoring, and improved postoperative care, mortality and morbidity has come down greatly. In contrast to previous decades, current microsurgical techniques are associated with lower postoperative morbidity and cranial neuropathies, and mortality rates as low as 1%.^{11,12}

Microsurgical resection offers the advantage of tissue diagnosis and possibility of cure. Tumors with demonstrated growth need to be resected early because the size of the tumor directly influences CN preservation; early surgery should be contemplated in patients with demonstrated tumor growth. Also for the considerably large tumors with or without symptoms but evidence of mass effect, surgical resection should be considered.

Kaye et al¹³ reported that because tumors tend to recur rapidly and reoperation is likely to increase CN deficits due to postoperative scarring, complete excision of a lesion is essential. In our series, complete tumor excision was performed in 41.6%; near-total excision was achieved in 50% of patients, and only subtotal decompression in 8.3% of patients. We are not able to comment on the growth rate of these tumors in our series maybe because no recurrence was noted up to the last follow-up in our series.

A grading scale according to imaging results is useful to guide surgical management for this rare type of tumor. Lesions involving the hypoglossal canal can be classified as described by Nonaka et al⁸: type A, intradural tumor; type B, transdural and extradural dumbbell-shaped tumors; and type C, extracranial base tumor.

In our series, the surgical approach was based on the extent of the tumor. Intradural tumors with a large posterior fossa component were managed with the retrosigmoid approach in nine cases with the patient in a lateral position. For transdural and extradural dumbbell-shaped tumors, the far lateral approach was performed in one case. The midline suboccipital approach was used in two cases for large tumors with caudal extension through the foramen magnum. There was no case of extracranial cervical extension of tumor in our series.

Combined approaches are favored for large extra- and intradural dumbbell-shaped lesions. For asymmetric dumbbell lesions in which only the larger lobule is symptomatic, the approach solely for the larger lesion can be selected, with close monitoring or radiosurgery considered for the smaller lobe.

The suboccipital, retrosigmoid, or petrosal approaches can be used in the resection of lower CN schwannomas limited to the posterior fossa.¹⁴ Transcranial dumbbell-shaped tumors require additional transcervical access.¹² Recent skull base techniques such as the far lateral, transcondylar, and supracondylar approaches have been advocated to access and manage lesions in this complex area.^{15–18} A 2011 article by Nonaka et al⁸ advocates the dorsolateral extreme lateral infrajugular transcondylar exposure (ELITE) approach for surgery of hypoglossal schwannomas of the intracranial type and the anterolateral ELITE approach for dumbbell-

shaped tumors. There are only two reported cases of hypoglossal nerve function improvement after tumor resection.^{19,20} In our series, none of the operated patient improved in hypoglossal function.

Kida et al²¹ and Pollock et al²² reported the results of radiosurgery for nonacoustic schwannomas. In our series good tumor control with five patients (71.4%) having their tumor size unchanged and two patients (28.5%) having reduced tumor size was seen.

Conclusion

Complete microsurgical resection is often associated with a high rate of morbidity. Subtotal and near-total resection followed by stereotactic radiosurgery or observation now offers an alternative approach.

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