Long-Term Outcomes after Surgical Treatment of Jugular Foramen Schwannoma

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ABSTRACT

The purpose of this article is to clarify long-term outcomes following surgery for jugular foramen schwannomas. Fifteen adult patients underwent surgery, predominantly via the lateral suboccipital approach, for jugular foramen schwannomas between December 1987 and May 2007. All information was collected retrospectively from patient medical records at a university hospital. The main outcome measures were tumor regrowth and IX-X nerve function at median 84.3 months (range, 12 to 166 months) following surgery. Near total removal was achieved in 10 patients, and the remaining 5 patients had subtotal removal. Tumor regrowth was observed in 9 of 15 patients at 6 to 89 months (mean, 31.2 months) after surgery. The actual tumor control rate was $70.0 \pm 12.8\%$ at 5 years. Fourteen of 15 patients exhibited IX-X nerve dysfunction immediately after surgery, but 8 of these 14 patients experienced amelioration of the symptoms at the final follow-up period. No patients required additional treatment. We found in our study that for jugular foramen schwannoma, tumor volume should be reduced as much as possible to avoid severe IX-X nerve damages postoperatively. If a small amount of tumor remains, long-term follow-up is necessary.

KEYWORDS: Jugular foramen schwannoma, lateral suboccipital approach, glossopharyngeal nerve, vagus nerve

Jugular foramen schwannomas arising from cranial nerves IX, X, and XI are relatively rare, slowly growing benign tumors that constitute ~2.9 to 4% of all intracranial schwannomas.^{1,2}

The ideal primary treatment for jugular foramen schwannomas is total surgical removal. However, management of these lesions is particularly challenging as a result of the complex anatomical

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location and potential postoperative complications. Although different surgical approaches have been proposed, new postoperative cranial nerve deficits are reported in 12 to 90% of the involved cranial nerves.²⁻⁶ We report the cases of 15 patients diagnosed with jugular foramen schwannomas who underwent surgery in the Department of Neurosurgery at the University of Niigata, and discuss tumor control rates and the function of cranial nerves IX and X during a long-term follow-up period after surgery.

PATIENTS AND METHODS

Subjects comprised 15 patients (9 men, 6 women) with jugular foramen schwannomas treated surgically at the University of Niigata between December 1987 and May 2007. Patients with type 2 neurofibromatosis were not included. All data were collected retrospectively from patient medical records.

Clinical presentation, radiographic assessment, cranial nerve involvement, surgical procedures, and follow-up outcomes were reviewed. All patients underwent gadolinium-enhanced magnetic resonance imaging (MRI). Tumor size was defined as the largest diameter of the intracranial and the extracranial lesion on MRI. This study used Kaye's modified classification based on the findings from MRI. Tumors that were primarily intracranial with only minor extension into bone were classified as type A. Tumors located mainly within bone with or without an intracranial component were type B. Tumors that were primarily extracranial with only minor extension into bone or the posterior fossa were type C. Type D (dumbbell-shaped) tumors comprised lesions with significant intra- and extracranial portions. Patients were followed by interviews, neurological examination, and MRI every 3 or 6 months for 1 year at the outpatient clinic. Thereafter, follow-up imaging was performed every 1 or 2 years. Median follow-up after tumor resection was 84.3 months (range, 12 to 166 months).

Table 1 Clinical Characteristics of 15 Patients with Jugular Foramen Schwannoma

Case	Age/Sex	Side	Tumor Size (Intracranial; mm)	Tumor Size (Extracranial; mm)	Kaye's Modified Classification*	Preoperative Signs
1	52/F	R	41	0	A	VIII, cerebellar ataxia, sensory impairment
2	67/M	R	26	20	D	VIII, IX-X, XI, cerebellar ataxia
3	38/M	L	0	36	C	IX-X, XII
4	31/M	R	32	0	A	VIII, IX-X, XI, cerebellar ataxia
5	38/M	R	44	35	D	VIII
6	48/M	L	45	8	A	V, VIII, XI, cerebellar ataxia
7	35/F	R	29	6	A	IX-X, XI
8	62/F	L	26	20	A	VIII
9	47/M	R	22	20	D	IX-X, XI, XII
10	44/F	R	26	19	D	IX-X, XI
11	61/F	R	30	10	A	VIII, IX-X
12	66/F	R	30	10	D	VIII, IX-X
13	45/M	L	25	10	D	IX-X
14	39/M	R	42	20	D	VIII
15	18/M	R	50	10	D	VIII, cerebellar ataxia, IICP

*For description of tumor types, see text in the Patients and Methods section.
R, right; L, left; IICP, increased intracranial pressure.

RESULTS

Patient Characteristics

Clinical data for patients are outlined in Table 1. Mean age at surgery was 46.1 ± 13.8 years (range, 18 to 67 years). Tumor was located on the right side in 11 patients and on the left side in 4. The most common presenting sign was hearing loss (67%), followed by glossopharyngeal and vagus nerve impairment (60%), accessory nerve impairment (40%), cerebellar ataxia (33%), and hypoglossal nerve palsy (13%). One patient (patient 6) displayed facial paresthesia at the time of presentation. Another patient (patient 15) presented with increased intracranial pressure signs such as headache and vomiting.

Radiological Findings

Mean intracranial tumor diameter was 31.2 ± 12.3 mm (range, 0 [localized extracranially] to 50 mm), and mean extracranial diameter was 14.9 ± 10.8 mm (range, 0 [localized intracranially]

to 36 mm). According to the classification of Kaye and Pellet,⁷ tumor was type D in 8 of 15 patients, type A in 6 patients, and type C in only 1 patient (patient 3) (Table 1).

Tumor Removal

A lateral suboccipital approach, in the lateral semi-prone position, was used for 12 patients (Table 2). A combined transjugular and suboccipital approach was used in 2 patients. Patient 3, with type C tumor, underwent surgery using an infralabyrinthine approach. Near-total removal (NTR), defined as capsule of tumor remaining, was achieved in 10 patients in this series. Subtotal removal (STR), defined as resection of >90% of the preoperative tumor mass, was achieved in five patients, with complete removal of the intracranial portion. Intraoperatively, the nerve of origin was identified as the glossopharyngeal nerve (nerve IX) in seven patients and the accessory nerve in one patient (nerve XI). In the remaining patients, tumor origin remained unknown (Table 2).

Table 2 Surgical Findings for 15 Patients with Jugular Foramen Schwannoma

Case	Age/Sex	Kaye's Modified Classification*	Approach	Removal	Origin
1	52/F	A	Lateral suboccipital	NTR	Unknown
2	67/M	D	Lateral suboccipital	NTR	Unknown
3	38/M	C	Infralabyrinthine	NTR	Unknown
4	31/M	A	Lateral suboccipital	NTR	Unknown
5	38/M	D	Lateral suboccipital	NTR	Unknown
6	48/M	A	Lateral suboccipital	NTR	IX
7	35/F	A	Lateral suboccipital	NTR	IX
8	62/F	A	Lateral suboccipital	NTR	IX
9	47/M	D	Lateral suboccipital + transjugular	NTR	XI
10	44/F	D	Lateral suboccipital	NTR	IX
11	61/F	A	Lateral suboccipital	STR	Unknown
12	66/F	D	Lateral suboccipital	STR	IX
13	45/M	D	Lateral suboccipital + transjugular	STR	IX
14	39/M	D	Lateral suboccipital	STR	Unknown
15	18/M	D	Lateral suboccipital	STR	IX

*For description of tumor types, see text in the Patients and Methods section.
NTR, near-total removal; STR, subtotal removal.

Function of IX-X Nerves

Six patients showed no preoperative cranial nerve X-XI signs (Table 3). In six of nine patients who preoperatively presented with IX-X nerve dysfunction, the degree of IX-X nerve palsy was mild and did not result in swallowing disturbance or hoarseness in daily life. All patients except one (patient 6) exhibited IX-X nerve dysfunction immediately after surgery. A greater deficit in IX-X nerves was recorded in 9 of 14 patients. Postoperative status of IX-X nerves compared with preoperative status was unchanged in 5 patients, worse in 4 patients, and better in only 1 patient (patient 3). However, with long-term follow-up, IX-X nerve dysfunction ameliorated compared with immediately after surgery in 9 of 14 patients (Table 3). No patients required postoperative nasogastric tube feeding or tracheostomy, and no patients needed vocal cord injections during follow-up. All patients achieved good activities of daily life by final follow-up.

Tumor Control

An increase in residual tumor volume was observed in 8 of 15 patients at a mean of 31.2 months (range, 6 to 89 months) after surgery (Table 3). Two of these eight patients (patients 14 and 15) underwent second surgery at 41 and 10 months after the initial surgery, respectively. One patient (patient 6) underwent second surgery at 72 months and gamma-knife surgery at 89 months. No signs of regrowth have been seen as of 123 months after the first surgery (Fig. 1). Another two patients (patients 9 and 10) underwent Novalis (BrainLAB, Heimstetten, Germany) stereotactic radiosurgery at 156 months and gamma-knife surgery at 92 months, respectively. The remaining three patients received no additional therapy because no signs or symptoms caused by tumor regrowth on MRI were seen at a mean of 67.3 ± 43.3 months (range, 12 to 155 months). The actual tumor control rate was $70.0 \pm 12.8\%$ at 5 years (Fig. 2). Log-rank analysis revealed that recurrence-free survival tended to be better with NTR than with

Table 3 Long-Term Outcomes for 15 Patients with Jugular Foramen Schwannoma

Case	Age/Sex	Removal	IX-X Signs*			Regrowth [†] (months)	Additional Therapy	Final Follow-up (months)
			Preop	Immediate	Final			
1	52/F	NTR	None	++	None	–	–	87
2	67/M	NTR	++	++	++	6	–	72
3	38/M	NTR	++	+	+	–	–	12
4	31/M	NTR	+	+	+	–	–	90
5	38/M	NTR	None	++	+	–	–	88
6	48/M	NTR	None	None	++	66	Reop (72 months), GKS (89 months)	123
7	35/F	NTR	+	++	+	–	–	66
8	62/F	NTR	None	+	+	22	–	155
9	47/M	NTR	++	++	++	72	Novalis (156 months)	157
10	44/F	NTR	+	+	None	89	GKS (92 months)	166
11	61/F	STR	+	++	None	65	–	66
12	66/F	STR	+	++	+	–	–	22
13	45/M	STR	+	++	+	–	–	15
14	39/M	STR	None	++	None	27	Reop (41 months)	134
15	18/M	STR	None	++	None	7	Reop (10 months)	12

*Degree of IX-X nerve dysfunction preoperatively, immediately after surgery, and at final follow-up.

[†]Period in months from first surgery to regrowth on MRI.

+, mild dysfunction; ++, moderate or severe dysfunction; NTR, near-total removal; STR, subtotal removal; Reop, reoperation; GKS, gamma-knife surgery.

Figure 1 Magnetic resonance imaging (MRI) for patient 6 at various stages. Left column: axial views. Right column: coronal views. (A) Preoperative MRI with gadolinium enhancement showing a large cystic schwannoma of the left jugular foramen. (B) Postoperative MRI after near-total removal of the tumor. (C) MRI at 67 months after initial surgery showing recurrence of intra- and extracranial tumor. The patient underwent second surgery at 72 months and gamma-knife surgery at 89 months. (D) Final MRI (123 months after initial surgery) showing no signs of regrowth.

Figure 2 Kaplan-Meier plot showing tumor control after surgery.

STR, although no significant difference was identified due to the small sample size ($P = 0.08$) (Fig. 3).

DISCUSSION

Surgical Approach

Despite advances in neuroimaging, microsurgical techniques, and modern skull base surgery, complete removal of jugular foramen schwannomas remains challenging. These tumors frequently involve adjacent structures, such as the jugular bulb, carotid artery, middle ear, petrous bone, infratemporal fossa, and posterior fossa. Many reports have described various approaches to these regions. In tumors located mainly in the cerebellopontine angle, a lateral suboccipital approach provides adequate exposure.² For dumbbell-shaped tumors, paracondylar,⁸ extreme lateral transcondylar transjugular,⁹ infralabyrinthine, and infratemporal-translabyrinthine⁶ approaches have been proposed in addition to the lateral suboccipital approach. Tumors mainly confined to the skull base and neck have been approached by cervical transmastoid,² infratemporal fossa,¹⁰ transcondylar, far-lateral,⁶ and transcervical approaches.¹¹ The present study exclusively used a lateral suboccipital approach because all patients except one (patient 3)

Figure 3 Kaplan-Meier plots for patients who underwent NTR and STR showing tumor control after surgery. NTR, near-total removal; STR, subtotal removal.

displayed tumors located predominantly in the posterior fossa. For tumors within the jugular foramen, subcapsular extirpation via the enlarged jugular foramen allowed NTR or STR of the tumor. In two patients (patients 9 and 13) with dumbbell-shaped tumors (type D), a transjugular approach combined with a lateral suboccipital approach was used for the intra- and extracranial lesions. A lateral suboccipital approach alone does not allow complete resection in patients with not only intracranial tumor, but also extracranial and intraforaminal tumor. We now routinely use an infralabyrinthine approach combined with a lateral suboccipital approach for dumbbell-shaped tumors.

Tumor Control

Many authors have reported lower recurrence rates (0 to 23%) in patients with jugular foramen schwannoma compared with our series (53%) (Table 4).^{1-3,11-14} The higher recurrence rate in this study is probably attributable to both the lower rate of patients who underwent total removal and the longer follow-up period after surgery. In all except two reports,^{1,11} mean follow-up periods were relatively short (22 to 37 months). Total tumor removal obviously results in no recurrence even over the long term. However, if NTR or STR is unavoidable due to tumor involvement of

Table 4 Microsurgical Resection of Jugular Foramen Schwannomas

Author, year	Number of Patients	Follow-up (months; mean)	Total Removal	Recurrence	Postoperative IX-X Signs
Kaye et al, ³ 1984	13	NR	92%	8%	100%
Tan et al, ¹ 1990	14	12–240 (70)	77%	23%	62%
Samii et al, ² 1995	16	12–42 (22)	100%	0%	81%
Mazzoni et al, ¹² 1997	19	6–108 (28)	100% (NTR)	0%	84%
Leonetti et al, ¹¹ 2001	13	24–108 (71)	77%	8%	77%
Kadri and Al-Mefty, ¹³ 2005	6	2–66 (33)	100%	0%	67%
Sanna et al, ¹⁴ 2006	22	3–168 (37)	95%	0%	95%
Present study	15	12–166 (84)	67% (NTR)	53%	93% (final follow-up, 67%)

NR, not reported; NTR, near-total removal.

important adjacent structures, long-term follow-up is necessary. Even patients who underwent intracapsular excision, corresponding to NTR in this study, experienced tumor recurrence at 5 to 15 years after initial surgery.¹ In four of eight patients with regrowth on MRI in this study, the MRI findings were only confirmed > 5 years after initial surgery. Jugular foramen schwannomas usually reach a considerable size before diagnosis due to a naturally slow growth rate.¹¹ A long time is thus generally required for the residual tumor to regrow to a considerable size.

Outcomes of IX-X Nerve Deficits

In patients with jugular foramen schwannomas, dysphagia and hoarseness caused by the IX-X nerve dysfunction are the most common symptoms. In previous reports, most patients experienced postoperative deficits of nerves IX-X (Table 4).^{1–3,11–14} Few patients have displayed improved symptoms immediately after surgery. Postoperative IX-X nerve function is generally unchanged or deteriorated compared with preoperative function. Likewise, all of our patients except one showed dysfunction of the IX-X nerves immediately after surgery. Nine patients experienced worsened function postoperatively. However, dysfunction of the IX-X nerves in many patients gradually improved to the level that the patients were unimpeded in daily life due to adaptation and functional compensation.

Many reports have described the need for tracheostomy, nasogastric feeding, or vocal cord injection following severe damage to the IX-X nerves.^{1–3,5,6,11,13,14} Complete tumor resection is likely to increase the risk of permanent severe disability in swallowing and vocalization. Conversely, the STR group tended to show a higher rate of recurrence than the NTR group in this study, although no significant differences were identified. The volume of residual tumor probably correlates with the possibility of tumor regrowth. Our findings suggest the merit of reducing the tumor volume as much as possible, to the extent where the patient does not experience severe swallowing disturbance or hoarseness postoperatively. Rather than total resection, NTR or STR as performed in this study may avoid severe deficits requiring additional treatments.

Radical surgery

Gamma-knife surgery has recently provided a high rate of tumor control, excellent rates of lower cranial nerve preservation, low morbidity, and no mortality in patients with jugular foramen schwannomas.¹⁵ The study found that tumor regressed in 17 patients, remained stable in 16, and progressed in only 2 patients at a mean of 83 months after treatment. Five- and 10-year actuarial control rates were 97% and 94%, respectively. Only one patient showed worsening of preexisting IX-X

nerve function. Improvement of lower cranial nerve function was observed in 21% of all patients. In terms of both long-term tumor control rate and preservation of cranial nerve function, gamma-knife surgery is obviously superior to microsurgery. However, radiosurgery is usually only indicated for patients with small or medium tumors. In addition, risks include development of long-term radiation-induced neoplasms or secondary transformation of an initially benign tumor after radiosurgery. Microsurgical resection could conceivably remain the first strategy, particularly in younger patients with large tumors. In the present series, three patients underwent stereotactic radiosurgery for recurrent tumor. Although follow-up was shorter, tumor volume in these patients has not increased since treatment. Stereotactic radiosurgery is likely to prove effective when the recurrent tumor is not particularly large, to minimize the possibility of deteriorated IX-X nerve dysfunction on second resection.

CONCLUSIONS

Completely total surgical resection for jugular foramen schwannomas obviously leads to high long-term tumor control rates. However, the risk of a new deficit or deterioration of IX-X nerve function may be unacceptably increased. Some patients are likely to experience severe swallowing disturbance and hoarseness requiring additional treatment. The ideal strategy may thus be to reduce tumor volume as much as possible, to the extent that the patient does not suffer from severe IX-X nerve damage postoperatively. If a small amount of tumor must be left, long-term follow-up is necessary. Second surgery or radiosurgery should be selected as treatment for recurrent tumor based on tumor size or type of extension.

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