Twelve Cases of Jugular Foramen Neurinoma

Abstract—Twelve patients with jugular foramen neurinoma were operated on at our clinic between 1974 and 1990. The initial signs and symptoms were variable; dysfunctions of the 7th, 8th, 9th, 10th, and 11th nerves were frequently observed. Among these, involvement of the eighth nerve was most frequent, and three patients were given a misdiagnosis of acoustic neurinoma. Computed tomography scan and magnetic resonance imaging were useful not only for the correct diagnosis, but also for planning the surgical treatment and postoperative follow-up. Surgical resection was accomplished with four different approaches: (1) suboccipital approach without opening the jugular foramen, (2) suboccipital approach with opening the jugular foramen, (3) suboccipital approach with opening the jugular foramen combined with infralabyrinthine approach, (4) infralabyrinthine approach. The surgical approach depended on the presence of intracranial tumor and on the extent of extracranial involvement. There was no operative mortality. Dysfunction of the 8th, 9th, 10th, or 11th nerve did not improve in any patient after tumor removal. In contrast, 12th nerve palsy improved in two of three patients after tumor removal. The nerve of origin was identified in five cases; those were from the ninth nerve in three and from the 11th nerve in two.

Jugular foramen neurinomas are relatively uncommon tumors and no large series exists in the literature. The initial signs and symptoms are variable and may not be associated with dysfunctions of lower cranial nerves.^{1–5} This variability of presenting symptoms frequently leads to an initial misdiagnosis for acoustic neurinoma.^{1,4–6} Recent advances in neuroradiologic diagnostic tools, such as computed tomographic (CT) scan and magnetic resonance imaging (MRI) have greatly improved our ability to differentiate these lesions accurately and to plan the surgical treatment.⁷ Recent advances in surgical approaches to the skull base have improved surgical results of this uncommon tumor.^{3,4,8–13}

We present 12 patients with neurinomas of the jugular foramen operated on at our clinic from 1974 to 1990 and discuss our surgical rationale in the management of this rare tumor.

CLINICAL MATERIALS

This series includes six men and six women (Table 1). The patients' ages at the time of admission ranged from 18 to 70 years, with a mean of 45.4 years. Eight patients had the tumors on the left side, and the other four, on the right side.

SIGNS AND SYMPTOMS

According to the growth patterns of tumors, our 12 cases were divided into three groups. Group A were patients who had tumors that were primarily intracranial with only minor extension into the extracranial space; group B were patients with tumors that expanded into the posterior fossa as well as into the extracranial space; and group C were patients who had tumors that were primarily extracranial with a minor extension into the posterior fossa (Figs. 1–3). The numbers of patients in each group were 5, 6, 1, respectively (Table 1).

As shown in Table 2, six patients (two in group A, four in group B) presented hearing difficulty as the initial symptom. Two patients in group B presented with tinnitus, and one patient in group A presented with vertigo. Thus, symptoms due to eighth cranial nerve dysfunction were present in 9 of 12 patients. Symptoms associated with lower cranial nerve dysfunction were seen only in two patients (one in group A and one in group C). One

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Case No.	Age, Sex*	Side	Preoperative Neurological Signs	Tumor Type†	Tumor Size (cm)	Nerve Origin
1	63, F	L	Cranial nerve: 7, 8, 9, 10; Ataxia, hemiparesis, choked disc	А	$3 \times 3 \times 3$	9
2	61, M	R	Cranial nerve: 7, 8, 9, 10, 11	А	$2 \times 2 \times 1.5$?
3	27, M	L	Cranial nerve: 5, 7, 8, 9	В	$4 \times 3 \times 2$?
4	28, M	R	Cranial nerve: 5, 7, 8, 9; Ataxia	А	$4 \times 3 \times 3$?
5	18, M	L	Cranial nerve: 6, 8; Ataxia	А	$5 \times 4 \times 3$	11
6	47, F	L	Cranial nerve: 7, 8, 9, 10, 11	А	$3 \times 3 \times 2$?
7	28, F	L	Cranial nerve: 8; Ataxia, choked disc	В	$5 \times 5 \times 4$	9
8	52, F	L	Cranial nerve: 5, 7, 8, 9, 10, 11, 12	В	$4.5 \times 3 \times 3$?
9	58, F	R	Cranial nerve: 9, 10, 11, 12	С	$3 \times 2 \times 1.5$	11
10	70, M	L	Cranial nerve: 8, 9, 10, 11; Ataxia, tetraparesis	В	$5 \times 4 \times 3$?
11	47, M	R	Cranial nerve: 8; Ataxia	В	$5 \times 4 \times 4$	9
12	46, F	L	Cranial nerve: 7, 8, 9, 10, 11, 12; Ataxia, hemiparesis	В	$6 \times 3 \times 3$?

Table 1. Clinical Data in 12 Cases of Jugular Foramen Neurinoma

*M: male; F: female; R: right; L: left.

⁺For a description of tumor types see text.

patient in group A experienced ataxic gait as an initial symptom.

Preoperative neurologic signs in patients from each tumor group are summarized in Table 3. In groups A and B, dysfunctions of the 7th, 8th, 9th, 10th, and 11th nerves were frequently observed. Among these, involvement of the eighth nerve was most frequent. In one case, impairment of the eighth nerve was the only presenting sign of cranial nerve involvement, although the patient had choked discs and ataxia. Of the 11 patients who showed eighth nerve dysfunction, nine had a certain degree of hearing loss, whereas the other two patients had vestibular nerve dysfunction without hearing loss.

Involvement of the 12th nerve was not observed in group A patients, even in cases with large tumors. Instead, it was observed in groups B and C. Therefore, 12th nerve palsy may be a sign of nerve compression in the extracranial space.

The one patient in group C presented typical signs of the Collet-Sicard's syndrome (9th, 10th, 11th, and 12th nerve palsies).¹⁴ Jugular foramen syndrome (Vernet's syndrome) is historically well known for these tumors.¹⁴ However, there was no patient in our 12 cases who presented only with the jugular foramen syndrome. All patients had some other cranial nerve palsies in addition to the 9th, 10th, and 11th nerve palsy.

RADIOLOGIC INVESTIGATIONS

Enlargement of jugular foramen was confirmed by plain skull films, tomography, or CT scan. Four of five



Figure 1. Computed tomography scans in a patient with group A tumor. Upper: Preoperative scans with contrast enhancement. Lower: Postoperative scans with contrast enhancement. This patient showed no marked abnormality of jugular foramen (arrow heads).



Figure 2. Computed tomography (CT) scans and magnetic resonance imaging (MRI) in a patient with group B tumor. Upper: Preoperative CT scans with contrast enhancement. Middle, left: Preoperative MRI, T_1 -weighted image. Middle, right: Preoperative MRI, T_2 -weighted image. Lower, left: Preoperative MRI, T_1 -weighted image with contrast enhancement. Lower, right: Postoperative MRI, T_1 -weighted image with contrast enhancement. CT scan clearly showed bony erosion of the jugular fossa. Both intradural and extracranial extension of the tumor was clearly demonstrated by MRI, especially contrast-enhanced MRI with Gd-DTPA.



Figure 3. Magnetic resonance imaging (MRI) and computed tomography (CT) scans in a patient with group C tumor. Upper: Preoperative MRI scans, T_1 -weighted images with contrast enhancement. Arrows indicate internal carotid artery. Arrowheads indicate the extracranial tumor. Lower: Postoperative CT scans without contrast enhancement. Arrowheads indicate a free fat graft filling the extradual defect.

Table 2. Initial Symptoms in Patients from Each Tumor Group						
	Group					
Symptom	A	В	C			
Hearing difficulty	2	4				
Tinnitus		2				
Vertigo	1					
Ataxic gait	1					
Hoarseness	1					
Dysphagia			1			

Table 3.	Preoperative Neurological Sign	IS
in Pat	ients from Each Tumor Group	

	Group				
Presenting Signs	Ā	В	<u>с</u>		
5th	1	2			
6th	1				
7th	4	3			
8th	5	6			
9th	4	4	1		
10th	3	3	1		
11th	2	3	1		
12th		2	1		
Hemiparesis	1	2			
Ataxia	3	4			
Choked disc	1	1			

patients in group A showed enlargement of both pars nervosa and pars vascularis. The other patient showed no marked abnormality of either pars nervosa or pars vascularis (Fig. 1). In group B, five patients showed enlargement of both pars nervosa and pars vascularis, whereas the other patient showed only enlargement of the pars nervosa. The one patient in group C showed enlargement of both pars nervosa and pars vascularis. Erosion or widening of the meatus acousticus was also noticed in two patients of group A and five patients of group B (Fig. 4).

Cerebral angiography was performed in all 12 cases. Although tumor stain was faintly observed in five cases, marked vascularity of the tumor was not observed in any of these cases. Such reduced vascularity was helpful in differentiating the tumor from glomus jugulare tumors.

CT scan and MRI were useful for evaluating extension of the tumor. CT scan clearly delineated destruction of the petrous bone (Figs. 2, 4). In the recent four cases MRI was taken. These tumors showed signal characteristics of low-signal intensity on T_1 -weighted image and high-signal intensity on T_2 -weighted image (Fig. 2). In two cases, contrast-enhanced MRI with gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA) was taken. In those two cases tumors were enhanced with Gd-DTPA (Figs. 2, 3).

The internal carotid artery is visualized as signal void on MRI. Therefore MRI was of great help in assessing the relationship between the tumor and the internal



Figure 4. Computed tomography scans showing the bony erosion of porus acoustics (left) and the enlargement of jugular foramen (right).

carotid artery in the region of the posterior petrous bone (Fig. 3). This information was helpful for avoiding damage to the internal carotid artery during the operation.

PREOPERATIVE DIAGNOSIS

Of our 12 cases, three (one in group A and two in group B) were misdiagnosed as acoustic neurinoma. In the case of group A, a correct diagnosis was made after operation. In the two cases of group B, even after the operation, correct diagnosis was not known until recurrence of the tumor, when the origin of the tumor was finally identified as the jugular foramen.

OPERATION AND RESULTS

In our 12 cases four different surgical approaches were undertaken: (1) suboccipital approach without opening the jugular foramen, (2) suboccipital approach with opening the jugular foramen, (3) suboccipital approach with opening the jugular foramen combined with infralabyrinthine approach,¹³ and (4) infralabyrinthine approach.¹² The second and third operative approaches will be briefly described.

Suboccipital Approach with Opening the Jugular Foramen

Patients were placed in the lateral position with the neck laterally flexed. A large curved incision was made, as shown in Figure 5, and the occipital bone, mastoid process, foramen magnum, and transverse process of the first cervical vertebrae were exposed, dividing the subcutane-



Figure 5. Schematic diagram showing skin incision and suboccipital craniectomy with opening of the jugular foramen.

ous tissues and muscles. After placing three or four burr holes on the suboccipital bone, posterior fossa craniectomy was performed with rongeurs. The bone covering the sigmoid sinus was removed with a high-speed air drill, and the sigmoid sinus was exposed down to the level of the jugular foramen. Next, the rectus capitis lateralis muscle and the atlanto-occipital ligament were detached from the jugular process of the suboccipital bone. With care not to damage the vertebral artery, the foramen magnum was opened and the posterior part of the occipital condyle was removed with a high-speed air drill. Finally, the posterior part of the jugular foramen was opened by removing the remaining jugular process of the suboccipital bone. By this procedure, extracranial tumor capsule continuing from the posterior fossa dura was exposed (Fig. 6). After the dura was opened as shown in Figure 6, cerebellum was gently retracted to the superomedial side and the cerebellopontine angle cistern was opened. Thus, both intracranial and extracranial portions of the tumor were ex-



Figure 6. Schematic diagram of the operative field following suboccipital craniectomy with opening of the jugular foramen. Extracranial tumor capsule continuing from the posterior fossa dura was exposed.

posed. In this procedure, neither mastoidectomy nor opening of the facial canal was performed. In cases with the tumor extending into the petrous bone, the bone lateral to the jugular bulb and the sigmoid sinus was further removed with a high-speed air drill. It provided enough space to remove the tumor extending into the petrous bone.

Suboccipital Approach with Opening the Jugular Foramen Combined with Infralabyrinthine Approach

This procedure was performed in cooperation with otolaryngologists. The sternocleidomastoid muscle was detached from the mastoid tip and the bone was removed with Gigli saw. Drilling away the mastoid, the facial nerve was identified just inferior to the horizontal semicircular canal and was followed down to the stylomastoid foramen. After complete decompression of the facial nerve, posterior fossa craniectomy and opening of the jugular foramen was performed as described before.

In four of five cases of group A, tumors were extensively removed by suboccipital approach without opening the jugular foramen (Table 4). In another case of group A, the tumor was totally removed by suboccipital approach with opening the jugular foramen. In this case, 9th, 10th, and 11th cranial nerves were resected together with the tumor.

In three of six cases of group B, tumors were first removed partially by suboccipital approach without opening the jugular foramen. These three patients had subsequent growth of the tumor and were reoperated on 3, 4, and 10 years later. In the two patients reoperated on 3 and 4 years after the first operation, the recurrent tumors were extensively removed by suboccipital approach with opening the jugular foramen. In one of them, 9th, 10th, and 11th nerves were resected with the tumor. In the third patient, operation was repeated with suboccipital approach without opening

Table 4.	Surgica	al Procedu	ure
Correlate	ed with	Tumor Ty	pe

Surgical Procedure	Group			
	A	В	С	
Suboccipital approach without opening the jugular foramen	4	3*		
Suboccipital approach with opening the jugular foramen	1	1 + (3)	1	
Suboccipital approach with opening the jugular foramen, combined with infralabyrinthine approach		1 + (1)		
Infralabyrinthine approach	5	<u> </u>	1	

*These patients had subsequent growth of the tumor and were reoperated on. Parentheses indicate surgical procedures of the latest operation for the recurrent tumors.

the jugular foramen; this patient had recurrence of the tumor 10 years after the second operation, and the third operation was performed via suboccipital approach with opening the jugular foramen. One patient in group B was operated on by suboccipital approach with opening of the jugular foramen allowing extensive removal of the tumor. There is no recurrence during the last 2 years of follow-up. In another case of group B, the tumor was initially removed by the infralabyrinthine approach. However, the patient had recurrence of the tumor 3 years later and was reoperated on by the suboccipital approach with opening of the jugular foramen combined with infralabyrinthine approach. The last patient of group B underwent suboccipital approach with opening the jugular foramen, combined with infralabyrinthine approach; no recurrence was observed during the last 6 years.

One patient in group C was operated on by suboccipital approach with opening the jugular foramen, and the tumor was extensively removed. During the last 4 years, there has been no recurrence.

RECURRENCE OF THE TUMOR

Recurrence of the tumor occurred after five operations in four patients. These four cases were all group B patients. Regarding the surgical procedures in these cases, suboccipital approach without opening the jugular foramen was used in four of the five operations, and infralabyrinthine approach in the other. The time until the recurrence of the tumor was 6 years on average.

ORIGIN OF THE TUMOR

In 5 of 12 cases, origins of the tumor were identified during the operation. Those include 9th nerve in three and 11th nerve in two (Table 1).

SURGICAL RESULTS

There was no operative mortality. Except for one patient, no complication was observed other than cranial nerve dysfunction. This one patient was group A and was operated on in 1974 by suboccipital approach without opening the jugular foramen. After the operation, this patient developed left hemiparesis, ataxia, and dysphagia.

As to the 9th, 10th, and 11th cranial nerve functions, impairments did not resolve in any patients after tumor removal (Table 5). At least some of those functions deteriorated further in three patients (cases 1, 4, 9) after the last operation. In the other four patients (cases 3, 5, 7, 11), dysfunction of some of those cranial nerves developed postoperatively. In three patients who had worsening or postoperative development of 11th cranial nerve palsy, the tumor was totally removed with the nerve. That was the reason for the deterioration. Also, postoperative dysfunction of the 10th nerve in case 3 resulted from resection of the nerve with the tumor.

In the other five patients (cases 2, 6, 8, 10, 12) who had moderate 9th, 10th, and 11th cranial nerve palsy preoperatively, the nerve functions were unchanged after the latest operation. Those five include one patient (case 6) in whom 9th, 10th, and 11th nerves were resected together with the tumor.

Before the latest procedure, three patients (cases 8, 9, 12) had 12th nerve palsy. After the operations, the 12th nerve function improved in two patients (cases 9, 12) and was unchanged in one patient (case 8). Of the other nine patients who did not have 12th nerve palsy before the operation, one patient (case 5) developed transient 12th nerve palsy after the operation.

Except for the one patient in group C (case 9), 11 patients had eighth nerve dysfunction before the most recent operations; of these, nine had hearing loss, which did not improve following the tumor removal. One patient

(case 3) who had moderate degree of hearing loss before the operation became deaf postoperatively. Mild temporary worsening occurred in the other patient (case 4). Of two patients (cases 5, 11) who had vestibular nerve dysfunction without hearing loss, moderate hearing loss occurred in one (case 11) after the operation.

Seven patients (cases 1, 2, 3, 4, 6, 8, 12) had mild facial palsy before the most recent operations; in five of those seven patients, the facial palsy was unchanged after the operation. In another two patients (cases 1, 4), it became worse. In two (cases 7, 11) of the other five patients without preoperative facial palsy, mild weakness developed transiently after the operation.

As to the other cranial nerve functions, sixth nerve palsy newly developed in three patients (cases 1, 4, 11) after the operation, although those were all transient. In one patient (case 5) who had sixth nerve palsy before the operation, it worsened transiently after the operation.

Impaired function of the fifth nerve that was observed in three patients (cases 3, 4, 8) before the operation did not obviously improve after the tumor removal.

DISCUSSION

Patients with neurinomas generally present paresis of the nerve of origin for the tumor early in the clinical course. In jugular foramen neurinomas, however, the initial signs and symptoms are variable and may not be associated with dysfunction of the lower cranial nerves.^{1–5} In our series, six patients presented hearing loss as the initial symptoms. Neurologic examinations on admission revealed eighth nerve involvement most frequently. Furthermore, the meatus acousticus and the jugular foramen were enlarged in seven patients. These features occasionally led to a misdiagnosis for acoustic neurinoma.^{1,4–6} In our 12 cases, three were misdiagnosed as acoustic neurinoma.

	Cranial Nerve*								
Case				8th					
No.	5th	6th	7th	Co.	Ve.	9th	10th	11th	12th
1		ND (T)	w (L)	u		w (L)	w (L)		
2			u	u		u	u	u	
3	u		u	w (L)		u†	ND (L)†	ND (L)†	
4	u	ND (T)	w (T)	w (T)		w (L)			
5		w (T)			i	ND (T)	ND (T)	ND (L)†	ND (T)
6			u	u		u†	u*	u†	
7			ND (T)	u		ND (L)	ND (L)	ND (L)	
8	u		u	u		u	u	u	u
9						u	u	w (L)†	i
10				u		u	u	u	
11		ND (T)	ND (T)	ND (L)	w (L)	ND (L)			
12			u	u		u	u	u	i

Table 5. Cranial Nerve Function Before and After the Operation

*Co.: cochlear nerve; Ve.: vestibular nerve; w: worsened; u: unchanged; i: improved; ND: newly developed; T: transient, L: longlasting CT scan and MRI were quite useful for the correct diagnosis. CT scan clearly showed bony erosion of the jugular fossa and posterior petrous bone. In some cases, however, the extent of the tumor could not be clearly visualized on CT scan because of bony artifacts. For visualizing the tumor, MRI was more helpful, as reported by Matsushima, et al.⁷ Both intracranial and extracranial extension of the tumor was more clearly demonstrated by MRI, especially contrast-enhanced MRI with Gd-DTPA. Furthermore, MRI was of great help in understanding the relationship between the tumor and the internal carotid artery in the region of posterior petrous bone. This information was helpful for avoiding damage to the internal carotid artery during the operation.

In consideration of the surgical indication, it is important to keep in mind the postoperative course of lower cranial nerve functions. Kaye et al.,⁴ who reported on 13 cases of jugular foramen neurinomas, stated that all but one patient had 9th, 10th, and 11th nerve dysfunction after operation. Horn et al⁵ have also stated that complete removal of the jugular foramen neurinomas is usually synonymous with paralysis of 9th, 10th, and 11th cranial nerves. In our 12 cases, dysfunction of the 9th, 10th, or 11th nerve did not improve in any patient after the tumor removal. At least some of the lower cranial nerves deteriorated in seven patients after the latest operations. Therefore one cannot expect to improve 9th, 10th, and 11th nerve function by tumor removal; the best surgical results may be to preserve the preoperative condition.

In contrast, 12th nerve palsy can be improved after operation. In the three cases with preoperative 12th nerve palsy, it improved in two after tumor removal.

It has been reported that hearing loss detected before the operation improved in a few cases following tumor removal.^{15,16} Our nine patients had certain degree of hearing loss before the latest operation, and it did not improve in any case following tumor removal.

Although these tumors may arise from any nerves passing through the jugular foramen, it is not always possible to identify the site of origin. In the literature³ the nerve of origin is described to be in order of 9th, 10th, and 11th nerve complex (that is, unknown) > 9th nerve > 10th nerve > 11th nerve. We could identify the nerve of origin in only 5 of 12 cases; those were from the ninth nerve in three and from the 11th nerve in two.

Surgery for group A tumors is not technically difficult. The tumor can be totally removed by suboccipital approach without opening the jugular foramen. By contrast, group B and group C tumors present technical difficulties. In those tumors, it is not possible to achieve total or subtotal removal of the tumor without opening the jugular foramen. In our cases, recurrence of the tumor was observed in four patients. These four cases were all group B tumors. In three of those four cases, it was attempted to remove the tumor by suboccipital approach without opening the jugular foramen. Presumably, the tumor extending into the posterior petrous bone or the extracranial space could not be removed. In another case, the tumor was first removed by the infralabyrinthine approach. In this case, the tumor around the jugular foramen and in the posterior fossa could not be removed. Those recurrent tumors were all successfully removed by opening the jugular foramen and posterior fossa dura.

Suboccipital approach with opening the jugular foramen combined with infralabyrinthine approach¹³ provides excellent exposure of the jugular bulb, the internal carotid artery, and the facial nerve in the region of posterior petrous bone, in addition to the intradural structures, and allows more complete removal of the group B and group C tumors. In this approach, however, there are a few disadvantages. This combined surgical procedure requires a team of neurosurgeons and otolaryngologists, and it takes longer. Isolating and transposing the facial nerve frequently produces transient facial nerve palsy.¹²

In the last five cases (group A one; group B three; and group C one) of our series, we employed suboccipital approach with opening of the jugular foramen, without combining infralabyrinthine approach. The tumors were successfully removed in all cases; postoperative morbidity resulting from lower cranial nerve dysfunction was not severe. No patient required tracheostomy. Dysphagia was minimal, and nasogastric alimentation was not required. In this approach, neither mastoidectomy nor isolation of the facial nerve in the bony canal was necessary. Therefore the duration of surgery was shortened. Furthermore, potential risk of damaging the internal carotid artery is small in this approach. After removing the extracranial portion intracapsularly, the internal carotid artery was easily found anterolateral to the tumor capsule. In the case where the tumor extended into the petrous bone, the bone lateral to the jugular bulb and the sigmoid sinus was further removed with a high-speed air drill. It provided enough space to remove the tumor extending into the petrous bone. By using this approach, we do not find it necessary to combine infralabyrinthine approach. However, our experience is still limited, and we have no experience of the surgery for large group C tumors extending and destroying bone of the infralabyrinthine and apical compartment of the temporal bone. In such cases, infratemporal fossa approach9,10 may be necessary for the radical removal of tumors.

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