Function-Preserving Multimodal Treatment for Jugular Foramen Meningiomas

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Abstract

Keywords

- jugular foramen meningiomas
- function preservation
- lower cranial nerve
- stereotactic radiosurgery
- long-standing tumor control

Objectives Despite being pathologically benign, jugular foramen meningioma (JFM) may be locally aggressive and spread in three compartments. Because of the complex anatomical location, radical removal of JFM usually causes serious morbidity through lower cranial nerve (LCN) deficits. To accomplish long-standing tumor control with good functional outcomes, we report function-preserving multimodal treatment (FMT) for JFM, comprising the combination of intradural tumor removal with the preservation of LCN function and stereotactic radiosurgery (RS) for the residual tumor.

Materials This study investigated six JFM patients (five women, one man). Preoperatively, five patients showed no LCN sign.

Results All patients underwent function-preserving retrosigmoid intradural tumor removal, and no patient developed new LCN deficit. Three patients underwent RS for the residual tumor at 8 to 12 months after surgery. After RS, all three tumors were controlled. No patients showed tumor recurrence or new LCN deficits in the follow-up period (2 months to 8 years).

Conclusion FMT for JFMs can accomplish long-standing tumor control with excellent functional outcomes.

Introduction

In the jugular foramen (JF), schwannomas and paragangliomas are the most frequent tumors.^{1–5} JF meningioma (JFM) arises from arachnoid-lining cells situated within the JF.⁶ Primary meningioma in the JF is very rare.⁷ Furthermore, despite being pathologically benign, JFM may be locally aggressive and spread into three compartments (intradural, intrapetrous, and extracranial). Because of the complex anatomical location,

radical removal of JFM usually causes serious morbidity in the form of lower cranial nerve (LCN) deficits. Management of this lesion is particularly challenging.

To accomplish long-standing tumor control with good functional outcomes, we report function-preserving multimodal treatment (FMT) for JFMs, comprising a combination of intradural tumor removal with the preservation of LCN function and stereotactic radiosurgery (RS) for the residual tumor.

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Patients and Methods

From April 2008 to November 2015, we encountered six JFM patients (five women, one man). Five of them showed no LCN signs preoperatively. Mean age was 54 years (range: 44–66 years). ►Table 1 summarizes the clinical symptoms, type of surgery, new postoperative dysfunction, postoperative radiation, and effect of radiation. The tumor volume was measured using noninvasive volumetric software (GammaPlan or SurgiPlan, Elekta AB). The volume measurements were made by two neurosurgeons. For RS, we used the multiple isocenter technique and set the optimal prescribed dose at 12 to 15 Gy directed to the 50% isodose line, changing this value according to individual situations such as tumor size.

Surgical Technique

All cases received function-preserving surgery for JFM, comprising removal of as much tumor as possible while preserving LCN function. A standard retrosigmoid approach was performed in all cases using a lazy S-shaped postauricular incision. The edge of the sigmoid sinus was slightly skeletonized. Intradural procedures of tumor removal were as follows: excision of tumor attachment to the petrous dura in the early stage, tumor debulking, and subsequent exposure of LCNs. The caudal part of the tumor was removed and then the petrosal vein and V/VII/VIII cranial nerves were exposed. In cases with intrameatal tumor extension, the internal auditory meatus was opened in 180 degrees of circumference, and the fundus side of cranial nerves VII and VIII were exposed. In the last stage of tumor removal, the LCNs were followed to the JF (►Fig. 1). In all cases, the facial and vagal nerve functions were monitored using the NIM Monitoring System (Medtronic); the vagal nerve function was monitored using an NIM EMG Endotracheal Tube (Medtronic). In cases with intrameatal tumor retention, the hearing was monitored using auditory brainstem response.

Fig. 1 The final view of tumor removal (right side).

Results

All patients underwent function-preserving retrosigmoid intradural tumor removal, and none developed any new postoperative LCN deficits. Three patients (cases 1, 4, and 5) underwent RS for the residual tumor at 8 to 12 months of surgery. RS achieved tumor shrinkage in two patients, and all three tumors were controlled after RS. None of the six patients showed either tumor recurrence or new LCN deficits in the follow-up period (range: 2 months to 8 years) (Table 1).

Illustrative Cases

Case 1
A 60-year-old woman visited the outpatient clinic with a 5-month history of hoarseness. Neurologic examination on admission showed LCN signs: hoarseness, dysphagia, and the atrophy of sternocleidomastoid muscle. Magnetic resonance imaging (MRI) with gadolinium (Gd) administration showed a cerebellopontine angle (CPA) tumor with a “dural tail” connecting to the JF, suggesting the diagnosis of meningioma

Table 1 Summary of the characteristics of six patients with JFM

Case, age, gender	Symptoms	Surgery	Postoperative new deficits	Stereotactic radiosurgery	Effect of radiosurgery	FU (years)
Case 1, 60, F	LCN sign (+): X/XI palsy	2008/April (TRFP)	None	2009/May (GK)	Shrinkage (67%)	8
Case 2, 44, F	LCN sign (–): gait disturb	2010/May (TRFP)	Transient hoarseness	None	Scheduled	6
Case 3, 66, F	LCN sign (–): hearing disturbance	2012/November (TRFP)	Transient hoarseness	None		3.5
Case 4, 66, F	LCN sign (–): headache	2013/April (TRFP)	None	2013/November (GK)	Shrinkage (41%)	3
Case 5, 45, F	LCN sign (–): headache	2013/August (TRFP)	None	2014/August (GK)	No remarkable change	3
Case 6, 44, M	LCN sign (–): hearing disturbance	2015/November (TRFP)	Transient slight swallowing dist.	None		<1

Abbreviations: F, female; FU, follow-up; GK, Gamma Knife; JFM, jugular foramen meningioma; LCN, lower cranial nerve; M, male; TRFP, tumor removal with function preservation.

Fig. 2 Pre- (left) and postoperative (right) T1-weighted magnetic resonance imaging (MRI) after gadolinium administration in case 1 with left-sided jugular foramen meningioma. The tumor was removed by function-preserving intradural tumor removal.

(► **Fig. 2**). Intradural tumor removal with the preservation of LCN function was performed. The postoperative course was uneventful. Ten months after tumor removal, the patient underwent Gamma Knife (Elekta AB) for the residual tumor. MRI showed a tumor shrinkage rate of 67% as of 8 years after RS, and her hoarseness subjectively ameliorated (► **Fig. 3**).

Case 4

A 66-year-old woman presented to the clinic complaining of headache and light-headedness. Neurologic examination showed no LCN palsy. MRI with Gd administration showed a CPA tumor with “dural tail” connecting to the JF, suggesting the diagnosis of meningioma (► **Fig. 4**). Intradural tumor removal with the preservation of LCN function was performed. No postoperative complications were encountered. Eight months after tumor removal, the patient underwent Gamma Knife surgery for the residual tumor, and a tumor shrinkage rate of 41% was observed 3 years after RS (► **Fig. 5**).

Discussion

Primary meningiomas of the JF are very rare, and less than 100 cases with primary JF meningioma were reported in the period from 1992 to 2007.⁷

JFMs arise from arachnoid-lining cells situated within the JF,⁶ but the exact origin of these lesions is confusing. Usually,

Fig. 3 Case 1: magnetic resonance imaging (MRI) at 8 years after radiosurgery shows tumor shrinkage.

Fig. 4 Preoperative T1-weighted magnetic resonance imaging (MRI) after gadolinium administration showing right-sided jugular foramen meningioma in case 4.

Fig. 5 Case 4: T1-weighted MRI after gadolinium administration prior to (left) and 3 years after (right) radiosurgery. The tumor shows a shrinkage rate of 41% after radiosurgery.

these tumors are large and involve the JF, jugular fossa, posterior fossa, and high cervical area. Arnautović and Al-Mefty⁸ use the term “jugular fossa meningiomas.” In many cases, it is quite difficult to know whether the exact origin of the lesion is in the jugular fossa or within the JF.

JFMs occur predominantly in women. Women comprised 83% of patients in our series, a proportion lower than that reported by Molony et al (100%),⁶ Arnautović and Al-Mefty (88%),⁸ or Ramnia et al (90%)⁴ but higher than those of other published cases (67%).^{7,9–14}

The mean age of our patients, 54 years, is higher than that listed in other reported cases (mean: 32–52 years).^{5–14}

Treatment of JFM includes observation, surgery, radiosurgery, and a combination of surgery and radiosurgery. Radiosurgery with Gamma Knife or LINAC (linear accelerator) may be an effective therapeutic modality in selected patients.^{5,8,15,16} In general, the indications for radiosurgery include advanced age, tumor volume < 20 mL, high operative risk, and refusal of surgery.^{16,17} On the other hand, planned partial resection and postoperative radiosurgery are yet to be evaluated in terms of the efficacy of long-term tumor control.^{7,10}

According to previous reports, radical removal of JFM is associated with serious morbidity in the form of LCN deficits.^{7,9,10,17} Most patients required temporary assistance with swallowing, airway protection, and phonation in the form of vocal cord medialization.^{7,9} Tracheostomy and/or gastrostomy should be performed as early as possible if persistent aspirations are present.^{7,17} This study investigated six JFM patients, all of whom underwent function-preserving retrosigmoid intradural tumor removal, and no patients developed new LCN deficits postoperatively. Three patients underwent RS for the residual tumor at 8 to 12 months after surgery. Our criteria for the postoperative RS include the following: (1) the postoperative intradural tumor remnant is confirmed by both intraoperative findings and postoperative MRI with Gd administration, (2) patients < 70 years of age, and (3) patient's consent for RS. Confirmation of the growth of postoperative remnant tumor using the MRI follow-up is not included in our criteria. After RS, all three tumors were controlled, and no patients showed tumor recurrence or new LCN deficits in the follow-up period (2 months to 8 years). These results indicate that FMT for JFM (i.e., intradural tumor removal with the preservation of LCN function followed by stereotactic radiosurgery for the residual tumor) can accomplish long-standing tumor control with excellent functional outcomes.

Conclusion

Despite being pathologically benign, JFM may be locally aggressive and spread in three compartments: intradural, intrapetrous, and extracranial. Because of the complex anatomical location, radical removal of JFM usually causes serious morbidity in the form of LCN deficits. This series showed that FMT for JFM (i.e., intradural tumor removal with the preservation of LCN function followed by stereotactic

radiosurgery for the residual tumor) can accomplish long-standing tumor control with excellent functional outcomes.

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