Long-Term Functional and Recurrence Outcomes of Surgically Treated Jugular Foramen Schwannomas: A 20-Year Experience

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OBJECTIVE: **To investigate the outcomes of jugular foramen schwannomas (JFSs) and to evaluate the risk factors for tumor recurrence and poor final outcomes.**

METHODS: **Between 1993 and 2013, 133 patients (68 female patients) with JFSs were surgically treated. Clinical charts were reviewed, and scheduled follow-up examinations were performed.**

RESULTS: **The average preoperative Karnofsky Performance Scale (KPS) score was 79.6. The JFSs were classified as follows: 65 cases, type A; 15 cases, type B; 5 cases, type C; and 48 cases, type D. Gross total resection was achieved in 107 (80.5%) patients. Transient and permanent morbidities affecting cranial nerves IX and X were 19.8% and 11.5%, respectively. After a mean follow-up duration of 108.0 months, 13 (9.9%) patients experienced recurrence. The most recent KPS scores averaged 83.7. Compared with the preoperative KPS score, the most recent KPS score was improved in 87 (65.4%) patients and stabilized in 29 (21.8%) patients. The presence of a solid tumor (hazard ratio [HR] = 5.815, $P = 0.010$), nontotal resection (HR = 4.613, $P = 0.007$), and pathologic mitoses (HR = 11.018, $P < 0.001$) were independent risk factors for tumor recurrence. Decreased preoperative KPS score (per 10 points) (odds ratio [OR] = 2.483, $P = 0.027$), a less soft tumor consistency (OR = 2.257, $P = 0.039$), and a solid tumor (OR = 3.755, $P = 0.041$) were risk factors for poor long-term outcomes.**

CONCLUSIONS: **Quality of life and preservation of neurologic function are the goals of surgical treatment of JFSs. Favorable long-term surgical outcomes for JFSs can be achieved. Morbidity of cranial nerves IX and X is significant, and patients with nontotal resection or pathologic mitosis should be followed closely.**

INTRODUCTION

Intracranial schwannomas account for 5%–10% of all primary central nervous system tumors[1,2]; only 2.9%–4% of these are jugular foramen schwannomas (JFSs).[3-6] These rare entities arise from cranial nerves (CNs) IX, X, or XI in the jugular foramen.[3,7] Because of the complex anatomic relationships of neurovascular structures surrounding the jugular foramen,[8-10] the limited surgical exposure and the restricted manipulative freedom of this region, and the susceptibility of the lower cranial nerves (LCNs) to injury, surgical management of JFSs is challenging and is accompanied by various morbidities. In the largest reported series of 81 cases by Sedney et al.,[11] transient morbidities were documented in 34.3% of patients, and permanent morbidities were documented in 22.3% of

Key words
- Jugular foramen
- Microsurgery
- Neurinoma
- Prognosis
- Schwannomas

Abbreviations and Acronyms
CI: Confidence interval
CN: Cranial nerve
GKS: Gamma Knife surgery
GTR: Gross total resection
HR: Hazard ratio
JFS: Jugular foramen schwannoma
KPS: Karnofsky Performance Scale
LCN: Lower cranial nerve
MRI: Magnetic resonance imaging
OR: Odds ratio
R/R: Recurrence/regrowth
WHO: World Health Organization

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patients. These neuropathies decrease the quality of life of patients and occasionally can be disastrous in elderly patients with no preoperative symptoms.[3] Before the series by Sedney et al.,[11] data of surgical experience and outcomes for JFSs were gleaned from small series, case reports, or series comprising diverse jugular foramen lesions.[3,12-18] The aim of the present study is to describe long-term outcomes, prognostic factors, and a proposed treatment strategy for JFSs.

MATERIALS AND METHODS

This study retrospectively reviewed 133 cases of JFSs, including 68 female patients (51.1%) and 65 male patients (48.9%) (female-to-male ratio approximately 1:1) who were surgically treated at Beijing Tiantan Hospital, Capital Medical University, from February 1993 to February 2013. Patients with neurofibromatosis type 2 were excluded. The Beijing Tiantan Hospital Research Ethics Committee approved the study. The Karnofsky Performance Scale (KPS) was used to evaluate preoperative and postoperative neurologic function. A dysphagia index was used to assess preoperative and postoperative swallowing function as follows: grade 0, normal; grade 1, problematic with compensation; grade 2, problematic with no compensation; and grade 3, unsafe swallowing.[19] For deaths resulting from surgical mortality or recurrence/regrowth (R/R) of lesions, the dysphagia index was ranked as 3. Preoperative and postoperative KPS scores and dysphagia grades were evaluated by 2 authors (X.J.-Z. and S.-Y.H.).

Preoperative conventional and contrast-enhanced magnetic resonance imaging (MRI) scans, with or without magnetic resonance venography, and computed tomography scans of the skull base were obtained routinely in all patients. Lesion size was evaluated as the lesion equivalent diameter $(abc)^{1/3}$, where a, b, and c represented the diameters as measured in axial, sagittal, and coronal magnetic resonance images. JFSs were classified into 4 types according to the Pellet-Kaye grading system: type A, primarily intracranial with minimal extension into the bone; type B, primarily within the bone with or without intracranial extension; type C, primarily extracranial with or without minor extension into the bone; and type D, dumbbell-shaped tumors with bone intracranial and extracranial components.[20-22]

Surgical Management

The surgical approach was selected according to lesion characteristics (lesion size, main location involved, classification, and extent of invasion), surgical goal, patient's wishes, preoperative neurologic status, and surgeon's preference. Intraoperative electrophysiologic monitoring was introduced at our institution in 1991 and was routinely used in the present series, including somatosensory evoked potentials, facial nerve electromyography, and monitoring of the LCNs. For type A lesions, a far lateral approach with or without a transcondylar approach was used; a retrosigmoid approach was considered for lesions with cerebellopontine angle extension, and in the early period of our series a suboccipital posterior midline approach was used in a few type A lesions that were lateral or dorsal to the brainstem. For type B and C lesions, a far lateral supracondylar/paracondylar approach (using a retroauricular arc-shaped incision) was preferred. Type D lesions manifested with various morphologic features making an individualized surgical approach necessary; retrosigmoid craniotomy was mandatory if the intracranial component was of considerable size. A far lateral supracondylar/paracondylar approach with a retrosigmoid craniotomy was the most common choice for type D lesions.

Intraoperative manipulation was meticulous and careful. After opening of the jugular foramen and exposure of the tumor, distinguishing the LCNs from the tumor was mandatory, especially for LCNs encased by tumor. The tumor mass was initially debulked by intracapsular piecemeal removal to reduce mass effect, and its capsule was then peeled from surrounding structures. After that, the intraosseous lesion was dissected meticulously from CNs IX–XI and the jugular bulb. Sharp/blunt dissection, rather than bipolar coagulation, was stressed because coagulation might cause thermal injury, vital vessel spasms, and microvessel occlusions. In patients with total loss of ipsilateral CN IX–X functions based on preoperative physical examination and intraoperative electrophysiologic monitoring, partial neurologic compensation usually developed, and radical resection was pursued with removal of the total tumor mass from surrounding structures with blunt/sharp dissection. Total resection was no longer attempted if there was tight adherence, injury to vital neurovascular structures, or obvious changes on neuromonitoring. In patients without dysphagia or with mild to moderate dysfunction, less aggressive resection and preservation of LCNs were priorities; dissection was performed using blunt separation as far as possible to avoid injury to LCNs; total removal was considered in cases of easy dissection with no or mild adhesion; residual tumor was left behind in cases with any changes on neuromonitoring or notable adhesion. Overall, the brainstem was spared regardless of preoperative status; sacrifice of neurologic function was not allowed to achieve radical removal. It was deemed preferable to peel tumor from CNs and brainstem only if no adherence was encountered or mild adherence could be separated effortlessly. It was considered prudent to remove tumors with tight adherence to CNs and brainstem that put neurologic function at risk. The surgical strategy could be adjusted in response to intraoperative findings.

Although computed tomography was routinely performed within 24 hours after surgery to check for surgical morbidities, a computed tomography scan was performed immediately after surgery if there was suspicion of hematoma or the patient was unresponsive. The extent of surgical resection was evaluated based on contrast MRI scans (**Figures 1–3**) as follows: gross total resection (GTR) (total removal without residual tumor on postoperative MRI), subtotal resection (95%–99% of the quantified lesion volume was removed), and partial resection (<95% of the lesion volume was removed). Postoperative MRI scans were ordinarily performed within 72 hours after surgery except in a few patients with extremely poor status (i.e., poor consciousness [a patient with a Glasgow Coma Scale less than 12], respiratory dysfunction, or severe dysphagia), in whom the MRI scans were delayed but were performed before discharge. The final diagnosis, World Health Organization (WHO) grade, and number of mitoses per 10 high-power fields (×400) were verified according to pathologic examination with Ki-67 immunohistochemical staining. Lesions were divided into 2 groups: no mitosis and ≥1 mitoses per 10 high-power fields.