Nonglomic Tumors of the Jugular Foramen: Differential Diagnosis and Prognostic Implications

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Key words
- Approach
- Infratemporal fossa
- Jugular foramen
- Nonglomic tumors
- Skull base surgery

Abbreviations and Acronyms
CN: Cranial nerve
CT: Computed tomography
JF: Jugular foramen
KPS: Karnofsky Performance Scale
MRI: Magnetic resonance imaging

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OBJECTIVE: **Tumors of the jugular foramen are notably rare, and the majority of them arise from the glomic tissue of the jugular vein. There are other tumors that do not originate from that tissue, and these are called the nonglomic tumors. This report includes a series of patients with nonglomic tumors of the jugular foramen to analyze their biological and radiological behavior and the clinical outcomes of patients.**

METHODS: **Patients with tumors of the jugular foramen other than chemodectomas were included for the present series. All of the patients were subjected to a protocol that included imaging studies and a complete clinical evaluation. Surgery was planned and performed by a multidisciplinary team using the following approaches: retrosigmoid, infratemporal fossa, and zygomatic-transmandibular. Depending on the precise diagnosis and surgical outcomes, radiotherapy or radiosurgery were indicated. The average follow-up period was 5 years.**

RESULTS: **Thirty patients with nonglomic tumors were included: 18 schwannomas, 6 meningiomas, 5 chordomas, and 1 metastatic carcinoma. The patients with chordomas had the most severe clinical manifestations, and the chordomas were the largest and most invasive tumors that destroyed the jugular foramen contour on imaging studies. Schwannomas presented a more benign clinical evolution and enlarged (but did not destroy) the jugular foramen contour. Two patients died (chordomas) during the follow-up because of tumor activity.**

CONCLUSIONS: **The most common nonglomic tumor of the jugular foramen was the schwannoma, which was the lesion with the best surgical prognosis. Chordoma is a rare and highly destructive tumor that has a notably high recurrence index.**

INTRODUCTION

The jugular foramen (JF) is a complex and deep structure located on the floor of the posterior fossa. The JF is divided by a fibrous, sometimes bony, septum into 2 segments: the *pars nervosa* and the *pars vascularis*. The *pars nervosa* is anteromedial, and the glossopharyngeal nerve (IX), the tympanic branch of the IX (Jacobson nerve), and the inferior petrosal sinus cross through this part. The *pars vascularis* is posterolateral, and the internal jugular vein, the vagus nerve (X), the accessory nerve (XI), and the auricular branch of the X (Arnold nerve) cross through it. Tumors that affect this region are rare. The majority of these tumors (60%–80% of the primary tumors) originate in the glomic tissue of the jugular vein (3, 6, 12, 14, 15). These tumors are known as chemodectomas, paragangliomas, or glomic tumors. The most common nonglomic lesions in this region are schwannomas, which are followed by meningiomas and bony tumors (2, 24). Less frequently, metastases, carcinomas, or sarcomas are found.

The signs and symptoms caused by tumors of the JF are similar among different tumor types (glomic and nonglomic) and in some cases, these tumors could be clinically confused with a cerebellopontine angle lesion. The clinical course is dominated by hearing loss, tinnitus, and/or alterations in the lower cranial nerves (CNs). Imaging studies are frequently nonspecific, and the precise diagnosis is commonly established after surgery.

The management of these lesions is complex and requires a multidisciplinary team that includes neurosurgery, otolaryngology, head and neck surgery, neurophysiology, and neuroradiology, among others. Because of the location and the size of these tumors, it is often necessary to carry out a combination of skull base approaches for their removal, which represents a nonnegligible risk for postoperative morbidity. Surgery of the JF is similar for glomic and nonglomic tumors, but paraganglioma cases typically are more challenging. Because of their high vascularity, glomic tumors require more complex preoperative procedures, such as angiography and selective embolization, and also require a wider exposure during their resection.

This work helps neurosurgeons to identify nonglomic tumors and to

differentiate these tumors from chemodectomas before treatment, allowing for better diagnostic and surgical planning and avoiding unnecessary surgical risks. In the international literature, we did not identify another series that includes the wide variety of diagnoses presented in this report.

PATIENTS AND METHODS

We carried out a cross-sectional, retrospective, retrolective study of a case series from 1998 to 2008. All the patients with lesions of the JF other than chemodectomas who underwent operation in the Center of Skull Base Surgery of the Neurosurgery Service of the Hospital de Especialidades del Centro Medico Nacional Siglo XXI in Mexico City were analyzed. All the patients were included in a protocol in which they underwent comprehensive clinical evaluation, computed tomography (CT), and magnetic resonance imaging (MRI).

The patients were subjected to surgery, and we planned the approach taking into account the specific tumor growth pattern. The portion of the tumor located on the posterior fossa was removed via a retrosigmoid approach, and the tumor component within the JF was removed via an infratemporal fossa approach (Fisch type "A"; **Figure 1**). For tumors presenting marked invasion of the infratemporal fossa, combinations of any of the previous approaches with zygomatic-transmandibular access were used (8) (**Figure 2**).

Depending on the complexity of the approach, the surgeries were planned in 1, 2, or 3 surgical stages. Total removal was considered when no residual tumor was observed macroscopically in the surgical bed and in control imaging studies carried out 6 weeks after the intervention. Any evidence of new tumor activity subsequent to that time was named as a recurrence. Subtotal resection was considered if residual tumor was left only on the JF. All the remaining resections were considered as partial. Depending on the histopathologic diagnosis and surgical outcome, the patients were submitted to adjuvant treatment, which was predominantly radiotherapy or radiosurgery. The average follow-up period was 5 years (range, 2–12 years). Clinical assessments and imaging studies were carried out at least twice per year.

The main goal of this series was to analyze the biological, clinical, and radiologic behavior of nonglomic tumors to propose guidelines to establish a more accurate differential diagnosis before surgery. Clinical outcomes were evaluated to establish some prognostic implications in these cases.

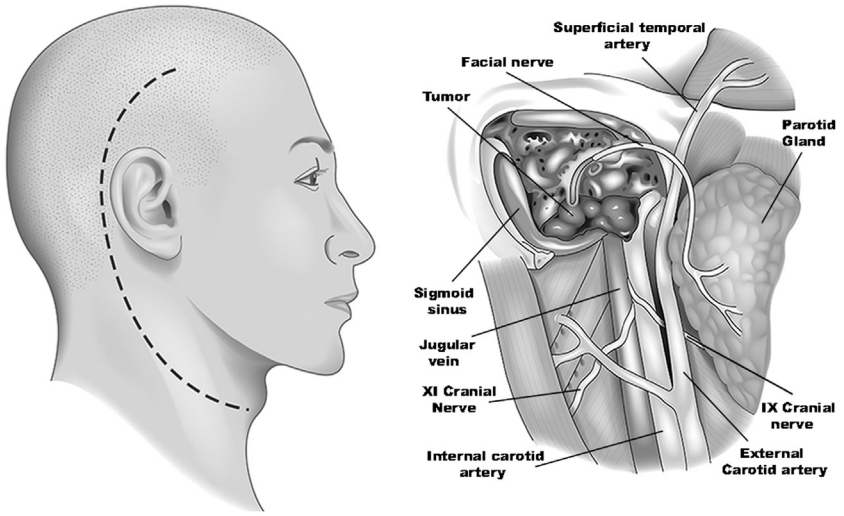

Figure 1. Infratemporal fossa approach. *Left*: Skin incision. *Right*: Tumor exposure. Note how anterior transposition of the facial nerve facilitates tumor excision. Carotid arteries, jugular vein, and lower cranial nerves are referred in the neck to assure vascular supply and nerve preservation.

Statistical Analysis

A descriptive analysis was performed regarding the signs, symptoms, imaging findings, surgical procedures, and evaluations of postoperative neurologic deficits. An inferential analysis was conducted regarding the diagnostic imaging findings using quantitative variables (tumor volumes and dimensions) and semiquantitative nominal categories. For enhancement on CT and MRI, the following ranking system was applied: 0 = null, 1 = mild, 2 = moderate, 3 = intense, and 4 = heterogeneous. The postoperative neurological status was evaluated in accordance with the deficit of at least 1 CN of the following 3 complexes:

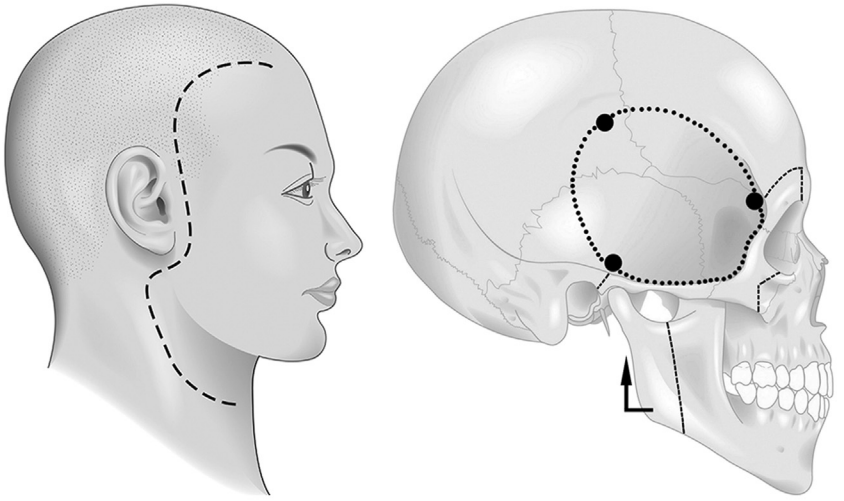

Figure 2. Zygomatic-transmandibular approach. *Left*: Skin incision. *Right*: Bone management. Access to infratemporal fossa is obtained through a pterional craniotomy and an orbito-zygomatic and mandibular osteotomy. Ascending ramus of the mandible is cut and rotated upward (*arrow*) using the temporomandibular joint as a hinge. This approach was used only in huge tumors with ample invasion to infratemporal fossa.