JUGULAR FORAMEN SCHWANNOMAS: DIAGNOSIS AND SUGGESTIONS FOR SURGICAL MANAGEMENT

Abstract—Schwannomas arising in the parapharyngeal space are rare lesions; however, those originating in the jugular foramen are even less common. Two cases, each with marked intra- and extracranial extensions, are discussed. Clinical presentation and preoperative evaluation emphasizing computerized tomographic and magnetic resonance imaging will be presented. An aggressive two-stage surgical approach consisting of a retrosigmoid craniectomy combined with infratemporal removal is advocated for those lesions arising within the jugular foramen.

Schwannomas arising in the parapharyngeal space are rare, with less than 150 cases being reported in the world literature.¹ Even less common are neuromas originating within the jugular foramen, of which 59 cases have been documented.² Preoperative evaluation of these lesions will be discussed with emphasis on computerized tomographic (CT) and magnetic resonance imaging (MRI). A two-stage surgical approach consisting of a retrosigmoid craniectomy combined with infratemporal removal is advocated for those lesions with both intra- and extracranial extension.

CASE REPORTS

Case 1

A 25-year-old man presented with right-sided hearing loss and tinnitus. The remainder of his cranial nerve and head and neck examination was normal. An audio-

gram demonstrated a 30 dB low frequency sensorineural hearing loss and auditory brainstem response results were consistent with retrocochlear disease on the right. Electronystagmography revealed a 30% right unilateral weakness. A CT scan (Fig. 1) demonstrated a 4 cm cerebellopontine angle lesion. An acoustic neuroma was suspected and the patient was taken to the operating room where removal of this tumor was accomplished by a translabyrinthine approach. At the time of surgery, the lesion was noted to extend inferiorly, being markedly adherent to cranial nerves IX, X, and XI. Pathologic examination revealed a schwannoma.

Two interval CT scans over the following year revealed no evidence of tumor. MRI (Fig. 2) and CT scans (Fig. 3) performed one and one half years subsequent to the first operation demonstrated recurrence of tumor in the cerebellopontine angle extending into the jugular foramen and neck. A two-staged procedure consisting of a retrosigmoid craniectomy to allow for intracranial removal followed by an infratemporal approach resulted in complete tumor removal. Postoperatively, the patient demon-

Figure 1. A preoperative axial CT scan with contrast in case 1 demonstrating a right cerebellopontine angle tumor.

Figure 3. A postoperative coronal CT scan with contrast in case 1 demonstrating recurrent tumor in the right cerebellopontine angle, jugular foramen, and neck.

Figure 2. A postoperative coronal MRI scan with gadolinium in case 1 demonstrating recurrent tumor in the right cerebellopontine angle, jugular foramen, and neck.

strated a right vocal cord paralysis and a reduced gag reflex.

Case 2

A 26-year-old woman complained of left-sided headaches partially relieved by aspirin. There were no other symptoms. Her head and neck examination was normal. An audiogram revealed normal hearing; however, an ABR

demonstrated prolonged interpeak latencies on the left. MRI revealed a 3×3 cm mass in the left cerebellopontine angle and posterior fossa (Fig. 4). The lesion was partially removed at another institution through a suboccipital approach. This procedure resulted in paralysis of cranial nerves IX, X, XI, and XII on the left.

A repeat CT scan 3 months postoperatively revealed tumor persistence with expansion of the jugular foramen and intracranial extension (Figs. 5, 6). An MRI scan demonstrated the tumor extending from the posterior fossa

Figure 4. A preoperative coronal T₁ weighted MRI scan in case 2 showing a mass in the left posterior fossa, cerebellopontine angle, and jugular foramen.

Figure 5. A postoperative axial CT scan with contrast in case 2 demonstrating recurrence of tumor in the left posterior fossa and jugular foramen.

Figure 6. A postoperative coronal CT scan with contrast in case 2 showing tumor recurrence in the posterior fossa, cerebellopontine angle, jugular foramen, and neck.

through the jugular foramen into the neck (Fig. 7). A two-stage procedure, utilizing a retrosigmoid craniectomy and later an infratemporal approach, led to complete tumor removal.

DISCUSSION

The jugular foramen, located between the temporal bone laterally and the occipital bone medially, is divided by a membranous or bony septum into two divisions. The larger posterior-lateral space transmits the internal jugular

Figure 7. A postoperative coronal T₁ weighted MRI in case 2 revealing tumor recurrence in the posterior fossa, cerebellopontine angle, jugular foramen, and neck.

vein and posterior meningeal artery, whereas through the smaller anterior-medial region pass the IXth, Xth, and XIth cranial nerves and the inferior petrosal sinus.

A variety of neoplasms can arise within or around the jugular foramen. These include glomus tumors, which are the most common tumor presenting in this anatomic location, followed by schwannomas, of which approximately 50% originate from the vagus nerve. In addition, meningiomas, chordomas, aneurysms of the carotid artery, carcinomas, sarcomas, and neuroblastomas have been found in this area.³ Schwannomas in this region are rare, with only 59 cases being reported in the world's literature.

The clinical presentation of neurilemmomas arising in the jugular foramen appears to fit into one of three categories. First, tumors in this area can cause Vernet's syndrome (jugular foramen syndrome).¹⁻⁴ This involves paralysis of cranial nerves IX, X, and XI due to tumor expansion within the foramen. In many reported cases, the nerve giving rise to the tumor is the first to be affected.⁵ A second mode of presentation mimics that of a glomus jugulare neoplasm. Jugular foramen schwannomas can present with pulsatile tinnitus, a reddish mass behind the tympanic membrane, and facial weakness.⁵ Finally, tumors arising within the foramen can give rise to the symptoms of hearing loss or disequilibrium and thus resemble an acoustic neuroma.^{2,4} These tumors can remain silent for a long period of time before causing symptoms

such as headache or hearing loss. As our two patients have demonstrated, these lesions can reach massive proportions before the onset of clinical symptoms.

Kinney et al² have described three patterns of growth of jugular foramen schwannomas, which these authors relate to the point of origin of the tumor within the jugular foramen. Lesions located distal in the canal will grow inferiorly, those located proximal will expand intracranially into the posterior fossa, and those tumors at the midpoint of the foramen can expand within the petrous apex and may reach the clivus.

The preoperative evaluation of these lesions has changed dramatically over the last several years with the introduction of sophisticated imaging techniques. CT and more recently MRI, in addition to angiography, have made the evaluation of jugular foramen schwannomas more precise. Typical CT findings of schwannomas arising in the jugular foramen include enlargement of the foramen in the anterior-medial direction as opposed to glomus tumors, which generally cause posterior-lateral erosion. Jugular foramen neuromas show more distinct margins on CT than is observed with glomus jugulare lesions. In addition, a mass in the posterior fossa or neck may be visualized (Fig. 6). MRI findings are even more dramatic. Generally, one can define the entire extent of the tumor, from the intracranial portion to that extending through the jugular foramen into the neck (Figs. 2, 7). One can usually separate a neuroma arising in the jugular foramen from one arising from the acoustic nerve with a properly targeted MRI. Angiography is very useful in the diagnosis of these lesions because one can observe external compression of the jugular vein as opposed to intraluminal involvement, as with glomus tumors. Additionally, chemodectomas will generally show a dramatic "tumor blush," whereas neuromas usually do not demonstrate this finding.

Traditionally, the surgical approach to these lesions has been through a suboccipital, retrosigmoid, translabyrinthine, or infratemporal approach. A combination of these techniques as a one-stage procedure has been advocated by Fisch.^{6,7} Due to these tumors being benign and generally slow growing, many surgeons have advocated watchful waiting. With advances in neuro-otologic surgery, the morbidity produced by tumor removal in and around the jugular foramen has been reduced. As is true for tumors elsewhere, the removal of smaller lesions produces less morbidity than the excision of larger tumors.

In order to maximize tumor exposure and removal and minimize morbidity, we suggest a two-stage approach. Initially, the intracranial component of the tumor can be removed through a retrosigmoid craniectomy. After outlining the sigmoid and transverse sinuses, the dura is opened and the cerebellum retracted posteriorly. Enough bone should be removed inferiorly to allow ample exposure of the IXth, Xth, and XIth cranial nerves as they leave the brainstem and enter the jugular foramen. While removing tumor from around the lower cranial nerves, recording the electrically evoked electromyography of the muscula-

ture associated with these nerves has proved beneficial in preserving their anatomic and functional integrity.

If the neuroma is large and hearing is markedly reduced, translabyrinthine removal can be combined with the above approach. The sigmoid sinus can be divided and ligated up to the vein of Labbe. Conversely, if hearing is useful, a hearing conservation procedure can be attempted. Three cases of hearing preservation following removal of jugular foramen schwannomas have been reported by Neely.⁸ Monitoring the auditory brainstem-evoked response intraoperatively appears to be of help during hearing preservation procedures.⁹

After the intracranial portion of the tumor has been resected, a second stage, infratemporal approach to the skull base, similar to the Fisch type A approach^{6,7} is advocated. Utilizing this approach, tumors within the jugular foramen and in the neck can be removed, allowing direct visualization of cranial nerves IX, X, XI, and XII. The jugular vein and bulb, generally found compressed by tumor, are removed to allow better visualization. For the same reason, the facial nerve is generally transposed anteriorly. As with glomus tumors, transposing the facial nerve anteriorly has met with minimal morbidity. Packing the wound with fat is advised to prevent cerebrospinal fluid leakage. Postoperatively, paralysis of the nerve of tumor origin is most often noted; however, paralysis of all cranial nerves within the jugular foramen may result secondary to tumor adhesiveness² or large tumor size.

SUMMARY

Jugular foramen schwannomas are rare lesions that can mimic an acoustic neuroma or glomus jugulare tumor. Clinically, they can be relatively silent or they can present with the jugular foramen syndrome. Preoperative evaluation should consist of CT, MRI, and angiographic studies. Combined evaluation allows more precise identification of tumor origin and extent. Surgical removal is recommended and a combined retrosigmoid/infratemporal fossa approach is advocated as a two-stage procedure to allow for complete tumor removal and prevent excessive morbidity. Allowing these lesions to reach massive proportions prior to recommending removal is argued against. Utilizing a two-stage neuro-otologic approach has reduced the morbidity related to surgical removal of these tumors, especially when these lesions are of a smaller size.

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