CASE REPORT

Glossopharyngeal Nerve Schwannoma: Report of a Case Operated on by the Far-Lateral Transcondylar Approach

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ABSTRACT—Complete resection with conservation of cranial nerves is the primary goal of contemporary surgery for lower cranial nerve tumors. We describe the case of a patient with a schwannoma of the left glossopharyngeal nerve, operated on in our Neurosurgical Unit. The far lateral approach combined with laminectomy of the posterior arch of C1 was done in two steps. The procedure allowed total tumor resection and was found to be better than classic unilateral suboccipital or combined supra- and infratentorial approaches. The advantages and disadvantages of the far lateral transcondylar approach, compared to the other more common approaches, are discussed.

Twenty-five percent of all schwannomas are found in the head and neck.¹ Intracranial schwannomas constitute approximately 8% of all primary brain tumors.² Schwannomas of the jugular foramen, arising from the ninth and 10th cranial nerves without associated neurofibromatosis are relatively uncommon³⁻⁴⁶ and account for only 2.9% of all intracranial schwannomas.⁴⁶ Less than 120 such cases have been reported^{11,14,21,32,41,46-48}; among these, some authors tend to include hypoglossal schwannomas, due to their anatomical and clinical similarity as well as difficulty in locating the nerve of origin.^{7,42,48-50}

Surgical lesions of the jugular foramen and nearby regions can be treated successfully by a combined far lateral approach to the craniocervical junction.

The far lateral approach to the inferior clivus and upper cervical region provides lateral extension to the unilateral suboccipital approach, which has been a neurosurgical standard for over a century.^{15,51-55}

We describe the case of a patient operated on by far lateral transcondylar approach for a schwannoma of the jugular foramen, originating from ninth cranial nerve,

and discuss the advantages of this approach for such lesions.

CASE REPORT

A 44-year-old man was referred to us in January 1998 with a 4-month history of swallowing difficulties, initially only with liquids and subsequently with solid food.

Craniospinal computed tomography (CT) and magnetic resonance imaging (MRI) detected an intra- and extracranial lesion localized in the left posterior cranial fossa about 6 cm in size: the lesion extended anteriorly as far as the posterior foramen lacerum and passed through it, presenting an extracranial portion in the region of the neck between the internal carotid artery and the jugular vein, which were closely involved. Neuroradiological investigation showed homogenous contrast enhancement and confirmed involvement of the above-mentioned structures, which were deformed and displaced by the tumor mass. The

patient underwent angiography, which demonstrated vascularization of the lesion, displacement of the vertebral artery, and venous reflux. MRI-angiography was performed to identify the relationship between the lesion and the vascular structures at the craniospinal junction (Fig. 1A and 1B).

The patient was intubated and positioned in right lateral decubitus. A left retromastoid lateral suboccipital incision was made: the lower edge of the incision was made in front of the sternocleidomastoid muscle, its central portion 10–12 cm from the ear, and the upper edge in the temporal region. After detaching the sternocleidomastoid and digastric muscles and removing three quarters of homolateral occipital condyle, an extensive craniectomy reaching inferiorly as far as the opening of the occipital foramen was made.

Subsequently, the dura was opened and the homolateral cerebellar lobe lifted up so that the intracranial intradural portion of the tumor could be removed as far as the jugular foramen and forward as far as the posterior foramen lacerum. The second surgical step was performed several days later to remove the extracranial portion after having removed the posterior arch of C1 and mobilized the vertebral artery.

Postoperative recovery was normal without neurological deficits; pharyngeal motility was preserved and there was gradual improvement of the dysphagia.

Postoperative MRI (Fig. 2A and 2B) confirmed the complete removal of tumor.

Histologic examination showed the tumor to be a schwannoma.

DISCUSSION

Schwannomas arising from cranial nerves and particularly those arising from the lower cranial nerves are rare. About 120 cases have been reported in the litera-

ture.^{11,14–19,21,32,41,46,47} These tumors arise from the 9th, 10th, and 11th cranial nerves. Using current neurodiagnostic techniques, preoperative diagnosis of jugular foramen schwannoma is usually almost certain. However, it is often impossible to determine the nerve of origin of tumor.⁷

Jugular foramen schwannomas are more commonly observed in females: the age at diagnosis ranges from 14 to 63 years (average 37 years).^{12,14–17,21–23,40,46}

Although the youngest patient in the literature was a 4-year-old boy,⁴¹ these tumors are infrequent in the first decade of life and in such cases should induce the suspicion of neurofibromatosis 2.²⁷

There seems to be no predilection for the left or the right side. Symptoms do not manifest until the tumor attains a fairly large size and, hence, vary from subtle to severe and may extend over years.

Tan et al⁴⁶ noted a length of symptoms ranging between 4 months and 10 years and Hakuba et al,¹⁴ observed a range from 3 months to 13 years, with an average of 2.7 years. Intracranial and bone tumors have the longest duration as opposed to extracranial tumors, which have a shorter duration.²¹ The origin as well as the size of tumor at the level of jugular foramen define clinical presentation and determine the surgical approach. If the tumor arises more proximally, it will have a mainly intracranial extension, presenting as a posterior fossa or cerebellopontine angle mass. More distal lesions manifest as extracranial, cervical, or skull base tumors. Tumors arising in the midregion will expand toward the temporal bone.^{10,27,56}

The early symptoms of presentation depend on the nerve of origin; however, this may not always be true for jugular foramen tumors because the compartment is so small that all cranial nerves may be involved at the same time. Horn et al¹⁷ observed that, although these tumors often present with a jugular foramen syndrome, palsies of the 9th, 10th, or 11th cranial nerves are not always present.^{20,40,43–45,48,57} Tan et al⁴⁶ reported involve-

Figure 1. Preoperative coronal (A) and sagittal (B) enhanced T1-weighted MRI scan (Philips, Gyroscan NT 15). The MR scan shows a mass presenting an homogeneous enhancement, involving the jugular foramen (A) and extending extracranially along the glossopharyngeal tract between the internal carotid artery and jugular vein (not shown). The brain stem result compressed and displaced (B) and the tumor extent along the inferior third of the length of the clivus (not shown).

Figure 2. Postoperative sagittal enhanced T1-weighted MRI scan (A) (Philips, Gyroscan NT 15) and axial CT scan of the skull base (B). MRI documents a complete removal of the tumor (A). CT scan shows a resection of the lateral mass of C1 together with the occipital condyle (B) allowing, with the far lateral access to the space anterior to the cervico-medullary junction, a better exposure of the region.

ment of the nerves of the jugular foramen in 8 of 14 patients.

In 1979, Hakuba et al¹⁴ reviewed the literature and found that at least half of patients had blurred vision with decreased visual acuity or diplopia, and dysphagia. Our patient only had dysphagia. Patients with larger tumors present 12th nerve involvement, increased intracranial pressure, papilledema, cerebellar mass, and/or brain stem signs.⁴⁶

A classification system helps to decide on the operative approach. At least three types of classification have been developed for jugular foramen schwannomas, depending on the location and extent of tumor: (1) Franklin et al¹² classified these tumors into Class A, B, and C, analogously to the categories used for glomus jugular tumors. (2) Kaye et al²¹ developed a similar but simpler grading system, as follows: (A) primarily intracranial tumors with minimal extension into the bone; B, tumors developing primarily within the bone, with or without an intracranial component; C, primarily extracranial tumors with only a minor extension into the bone or into the posterior fossa. Pellet et al³⁸ added a fourth type, D, which is a saddlebag-shaped type B tumor with intracranial and extracranial components linked via the jugular foramen. (3) Samii et al⁵³ classified tumor extension in relation to the radiological and surgical features into type A, B, C, and D, as follows: A, tumors primarily located at the level of the cerebellopontine angle with minimal enlargement of the jugular foramen; B, tumors primarily in the jugular foramen with intracranial extension; C, primarily extracranial tu-

mors with extension into the jugular foramen; and D, dumbbell-shaped tumors with both intra- and extracranial components.

Jugular foramen schwannomas are sometimes difficult to differentiate from acoustic tumors. Patients frequently present with seventh and eighth cranial nerve deficits by virtue of the proximity of the jugular foramen to the internal acoustic canal. This finding has been observed more frequently in cases of type A tumors by Kaye,²¹ whereas patients with type B tumors more often exhibit tinnitus, deafness, middle ear mass, or involvement of the hypoglossal nerve.²¹ There is no universal agreement regarding the position of cranial nerves in the jugular foramen. Arenberg and McCreary⁵⁷ and Gejrot⁵⁸ believed that the 9th, 10th, and 11th nerves are located in the pars nervosa, but Mountjoy³¹ and Bossy⁴ placed there only the 10th and 11th.

There is a common dural sheath for the 10th and 11th cranial nerves. The glossopharyngeal nerve has a separate dural covering as it passes through the foramen. The meningeal branches of the occipital and ascending pharyngeal arteries pass through the intermediate portion and are responsible for vascular supply to tumors in this region.

A variety of tumors arising from the structures within or adjacent to the jugular foramen are encountered in the cerebellopontine angle. Of great importance in differential diagnosis of jugular foramen schwannomas are acoustic schwannoma and glomus jugulare tumor. However, other tumors that involve the cerebellopontine angle, namely meningiomas, choroid plexus

papillomas, chordomas, exophytic pontine gliomas, chondromas, cerebellar hemangioblastomas, myxofibrosarcomas, epidermoid cysts, and carcinomas of the tympanic cavity, have been taken into consideration.^{3,5,9,15,16,45,46,50,58-60,61}

Plain x-ray films, which were essential until a decade ago, have become obsolete since the introduction of high-resolution CT (with soft-tissue and bone algorithms), and MRI.^{29,50,62} In Samii's type B tumors, considerable bone destruction was noted. In case of glomus jugulare tumors, irregular bone destruction of the jugular foramen is frequently identified.

All of these findings are better appreciated on high-resolution CT scans with bone windows. On CT scans without contrast enhancement, the tumor appears isodense or slightly hyperdense, and shows moderate enhancement after administration of contrast medium.⁴⁹ This is in contrast with meningiomas and glomus tumors, which show a marked enhancement. If the tumor is of considerable size, a fourth ventricular displacement is seen.³²

MRI and MR angiography have revolutionized the diagnosis of these tumors. The soft-tissue details, vascular supply, relationship with major vessels, as well as the patency of the jugular bulb are well appreciated on MRI with gadolinium enhancement and MR angiography. On T1-weighted images, these tumors are isointense or hypointense, becoming hyperintense on T2-weighted images.²⁹ They show significant enhancement after intravenous gadolinium administration. Absence of a tumor tail within the acoustic meatus usually differentiates them from acoustic schwannomas. Enhancement is considerably greater in meningiomas, whereas signal-void areas, indicating vascularity, are more prominent in glomus jugular tumors.^{29,30,32,62}

Conventional angiography cannot always be dispensed, because MR angiography is not yet fully developed. Occasional tumors of the jugular foramen have been described as being moderately vascularized,²¹ but not as much as the highly vascular glomus jugular tumors. Various changes are noted on angiography, including ventral convex displacement of the cervical portion of the internal carotid artery, displacement of the PICA to the opposite side, displacement of the SCA backward, and displacement of the tonsillar circulation.^{14,45,46,63,64} In larger tumors, compression or obstruction of the jugular bulb is often seen.⁵⁷ Sometimes the tumor can mimic a brain stem tumor on angiography.²⁸ Angiography is also useful for performing embolization if the tumor is highly vascularized.⁹

There is not a wide consensus regarding the management schwannomas involving the jugular foramen.⁶⁵ Mazzoni et al⁶⁵ reported 19 patients operated on and one case managed by "watchful expectancy." Fifteen cases were operated on by petro-occipital trans-sigmoid approach with or without labyrinthectomy, 2 by the in-

fratemporal fossa approach, 1 by the modified trans-cochlear approach, and 1 by the jugulo-petrosectomy approach. The petro-occipital trans-sigmoid approach allowed a single-stage, total tumor removal with preservation of the facial nerve and of middle and inner ear functions.

In 1978, Fisch⁶⁶ improved Gallard and Rebattu's cervicomastoid approach, calling it the "infratemporal approach." Various approaches have been described to resect these tumors depending on their location and extension. Tumors confined to the upper cervical soft tissues are frequently dealt with using a conventional lateral neck approach⁶⁷; occasionally, a modified incision combined with parotidectomy and cervical extension is performed if the tumor reaches the jugular foramen. The latter approach helps to mobilize the facial nerve.¹² These tumors can also be managed through a mastoid-hypotympanic neck approach used for glomus jugulare tumors.^{7,8} In type A tumors, a suboccipital craniectomy gives good exposure. In type C ones, the "infratemporal approach" described by Fisch⁶⁶ seems to be indicated because it provides direct access to the jugular foramen and neck and allows good control of neighboring major blood vessels.^{66,68} If the intradural tumor extension is more than 2 cm, this approach does not offer a full view of brain stem and cerebellum on which the tumor encroaches, and hence, in these cases, the type A infratemporal approach is used with a suboccipital craniectomy.¹² In type B tumors, suboccipital craniectomy with either a translabyrinthine transcochlear or an infralabyrinthine (retrolabyrinthine) approach has been advocated, depending on the function of the eighth nerve. Pellet et al³⁸ described the widened transcochlear approach for voluminous type B tumors with both intra- and extradural extension, their so-called "type D" tumors. In Samii's study, all the type A tumors were operated on through a retromastoid craniectomy and type B, C, and D tumors through a single-stage cervical approach combined with mastoidectomy, with the following advantages: (1) the facial nerve is left in its bone canal; (2) drilling of the petrous bone inferior to labyrinth and cochlea allows preservation of hearing and vestibular functions; (3) in case of injury of the spinal accessory nerve during cervical tumor resection, reconstruction can be performed using a nerve graft; and (4) mobilizing the sigmoid and transverse sinuses and the jugular bulb medially, a better exposure is obtained.⁵³

Tan⁴⁶ operated on 14 glossopharyngeal schwannomas: of these, one presented mainly in the neck, but the remaining 13 were predominantly intracranial.

Nine tumors were resected through a posterior fossa approach, three required a combined posterior fossa and neck dissection, and one a combined supratentorial and infratentorial craniotomy.

The basic far lateral approach without drilling of the occipital condyle may be all that is required to reach

some lesions located along the anterolateral margin of the foramen magnum. However, it also provides a route through which the transcondylar, supracondylar, and paracondylar approaches and several modifications of them can be completed. The transcondylar exposures can be separated into various subtypes^{50,69}: a variant is the atlantoccipital transarticular approach, in which the adjacent posterior portions of the occipital condyle and the superior articular facet of C1 are removed to facilitate a circular dura incision and to permit a mobilization of the VA with the surrounding cuff of dura. A more extensive removal of articular surfaces and condyles can be performed to gain access to extradural lesions situated along the anterior and lateral margins of the foramen magnum. Another subtype, the atlanto-occipital transcondylar approach, is directed above the atlanto-occipital joint through the occipital condyle and below the hypoglossal canal to access the lower and lateral side of clivus and the area in front of the medulla. The supracondylar approach directed above the occipital condyle can also be used, depending on the anatomy to be exposed.

In the transtubercular variant of the supracondylar approach, the prominence of the jugular tubercle, which blocks access to the area in front of glossopharyngeal, vagus, and accessory nerves, is removed with the aim of increasing visualization of the area in front of the brain stem and of exposing the origin of the PICA, which arises from the distal portion of VA near the midline.

The paracondylar approach also has several variants. In the transjugular variant, exposure is directed laterally to the condyle through the jugular process of the occipital bone and to the posterior surface of the jugular bulb. This approach can also be extended lateral to the jugular foramen into the posterior aspect of the mastoid to gain access to the mastoid segment of facial nerve and the stylomastoid foramen.⁶⁹

The far-lateral approach in which exposure is carried up to, but does not include, the posterior margin of the occipital condyle may be selected for the lesions located along the lateral or anterolateral aspect of foramen magnum. It is frequently necessary to remove a small portion of the occipital condyle and/or the superior articular facet of the atlas, if there is the need to complete a circumferential dural incision around the site at which the artery penetrates the dura. The artery can be displaced for access to lesions that are located ventral to it and in front of the cervicomedullary junction.⁶⁹

In our patient the surgical approach was a far lateral with partial removal of the homolateral occipital condyle. The lesion was large (6–8 cm) and was localized on the antero-lateral face of the foramen magnum straddling the posterior and middle cranial fossae, around the tuberculum jugulare and invading the nearby foramen jugulare.

This approach was preferred in view of the intra- and extracranial extension of tumor (dumb-bell-shaped; group D according to Samii's classification) and its close proximity to the internal carotid artery anteriorly, to the internal jugular vein laterally, to the brain stem medially and to the cerebellum posteriorly, as well as to the basilar artery in the intra-extracranial passage and the arch of C1 inferolaterally. Similarly to other authors,^{8–10,70,71} we preferred the park-bench position, which offers the main advantage of avoiding air embolism. According to other authors,^{10,39,53,59,67} it permits a wider angle of view that is associated with a less distension of venous plexus, even if the rich net of veins around cervical muscles and VA offers the risk of air embolism. The risk becomes greater if condylar emissary vein, hypoglossal venous plexus, sigmoid sinus, jugular bulb, and IJV are exposed.

We performed an "inverted horseshoe" skin incision, with the longer limb located laterally and with most of the cervical and suboccipital muscles reflected medially, as advocated by the de Oliveira et al⁵⁹ and by Babu et al.⁷⁰

In fact, it moves the bulk of muscular mass away from the direction of surgical approach. The longer limb located at the midline, with all cervical and suboccipital muscles reflected laterally, has also been recommended.^{54,72}

Anatomically, the horseshoe skin incision and muscular dissection, layer by layer with muscles reflected medially, allows additional space for the lateral approach and offers the best preservation of muscular landmarks such as the suboccipital triangle and levator scapulae (for locating VA) and the rectus capitis lateralis (for locating the posterior portion of jugular bulb).

CONCLUSION

For surgical treatment of intra- and extradural vascular and tumoral lesions of jugular foramen, the far-lateral approach and its variants, afford good visualization of all vascular and nervous structures contained in this region.

Resection of the occipital bone (articular condyle and antero-lateral portion) allows removal of any nervous structures (glossopharyngeal, vagus, accessory, hypoglossal nerves, etc.).

By extending this approach (removal of the posterior arch C1), it is possible to identify and mobilize the vertebral artery in the intra- and extracranial passage that provides wider dural opening and a larger surfaces for surgical access to intradural vascular or tumoral lesions localized antero-laterally to the brain stem in general and, as in our case, to dumb-bell-shaped extra- and intradural tumors of the low cranial nerves.

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