INTRODUCTION

Glomus jugulare tumors consist in rare, vascularized masses, belonging to the paraganglia tumors, considered a challenging pathology to treat. Approximately 80% of the paraganglia tissue is located in the suprarenal glands. The remaining 20% of the respective tissue has close association with autonomic and some cranial nerves. Although these tumors have shown in histopathological studies to present neurosecretory granules, they are usually nonfunctional.

Paragangliomas are classified in four types of tumors according to their origin, such as “glomus caroticum” from carotid artery baroreceptors, “glomus jugulare” from internal jugular vein (IJV), “glomus vagale” from along the vagal nerve, and “glomus tympanicum” from the tympanic cavity (Table 1). Believed to be originated from the glomus bodies, this tumor usually arise in the region of the Jugular bulb—because of in that region is found high number of glomus bodies. Glomus tumors can also arise in this region along parasympathetic nerves, especially IX and X cranial nerves (Jacobson’s and Arnold’s tympanic branches) (Table 1).

Considering the wide spectra of growing and presentation, these tumors can have similar presentation despite the initial location of the tumor. Sometimes the initial presentation or the progression of the deficits help us to characterize the initial site of the lesion.

In 1840, Valentine described for the first time the glomus tissue, referring to a ganglion as a small cellular formation close to the origin of the tympanic nerve. Guild coined the term “glomus jugularis” or “jugular body” to describe a paraganglionic tissue composed of capillary or precapillary vessels interspersed with numerous epithelioid cells. Indistinguishable from carotid bodies, the respective relationship with glomus tissue was first described by Krause in 1878.

The exact function of the glomus body remains unknown and its similarities with carotid body have suggested a chemoreceptor role related to hypoxia, hypercarbia and acidosis. Speculations about its function consist in the regulation of the microcirculatory blood flow or blood gas homeostasis and pressure in the middle ear.

SPECIFIC PREOPERATIVE EVALUATION

Epidemiology

Age presentation of the glomus tumors ranges from the 2nd to the 9th decade, although most tumors manifest during the middle age, affecting 6 times more women than men. Familial glomus has been described in literature, varying from a minority of cases report to 50% in some series.

Glomus tumors are usually benign, slow-growing lesions, locally invasive, and highly vascularized. In approximately 1–3% of the cases, these tumors present an aggressive form and growth, metastasizing to regional lymph nodes or to distant sites. These tumors might be also bilateral and associated with other neoplasm lesions, especially chemodectomas. Abdominal computed tomography (CT) scan might be useful in demonstrating the occurrence of concomitant pathologies such as adrenal tumors and glomus tumors.

Approximately 4% of the respective glomus tumors produces catecholamines and manifests symptoms such as blood pressure fluctuations during surgical intervention. Due to that fact, a 24-hour urinary study to screen vanillylmandelic acid, metanephrine, free catecholamines and 5-hydroxyindoleacetic acid may help to predict surgical behavior of the blood pressure in those patients.

Diagnosis

Imaging studies diagnosis involves CT (Computed tomography) scan, MRI and angiographic studies. Typical aspect of the lesion by angiography demonstrates a great blush
contrast, located at the site of glomus cells proliferation (Fig. 1). This respective vascular characteristic shows the difficult management of the lesions because of rich vascularization. CT scan represents the best imaging study to demonstrate bone lesions compromised by the tumor (Fig. 2). MRI may also show a hypo-isointense lesion that enhances widely with paramagnetic contrast, correlating to the vascular aspect of the lesion (Figs 3A and B).

Differential diagnosis must be considered when glomus tumors are suspected, such as schwannomas, meningiomas, chemodectomas, chordomas, chondrosarcomas, myxomas and epidermoid cysts.10

Clinical Aspects
The usual clinical presentation of the glomus tumors comprises of progressive unilateral hearing loss and pulsatile tinnitus, secondary to the mesotympanum invasion and rich vascularity of the lesion. Vertigo symptoms are explained once the labyrinth is invaded by the tumor. Other manifestations depend upon the tumor rate of growth and the extensiveness of the lesion, knowing the possibility to compromise cranial nerves dysfunction (VII, VIII, IX, X, XI, and XII), Horner’s syndrome, petrous bone posterior syndrome (V and VI), cavernous sinus invasion, brainstem compression, and hydrocephalus.

The senior author reported in his series hearing loss as the most common symptom, compromising about 88% of the cases, followed by dysphagia and dysphonia in about 50% of the patients.29

Recently, Cheesman et al. presented a protocol management that has been made based on a 30-year experience of 134 glomus jugulare tumors.11 The current protocol involves a preoperative assessment of swallowing. Considering that low cranial nerves palsies have great morbidity, it is considered to be important to know ahead the management of such deficits. The author mentioned describes that after jugular foramen surgical procedure, patients undergo further examination using fiber optic endoscopic evaluation of swallowing (FEES), video fluoroscopy and manometry studies. Those patients, presenting prolonged or poorly compensated dysphagia, are offered rehabilitation surgery.11 Tracheostomy and gastrostomy must also be considered in cases presenting lower cranial nerves deficits and prolonged intubation. Sometimes, the aggressive management of these deficits, considering the previous or postoperative deficits, can be the best option for the outcome. This management can help to avoid future or possible complications.

Surgical Anatomy
Comprising a complex region, the jugular foramen involves nerves, sinus cavities, dural folds, and osseous structures. Not only this respective complex region, but also the difficult approach to this area converts the foramen’s pathologies into a real challenge for the neurosurgeon.

The jugular foramen varies in size and shape for each person and in the same person, it can vary according to the
left or right side. It is surrounded by important structures: carotid artery (anteriorly) facial nerve (laterally), hypoglossal nerve (medially), and vertebral artery (inferiorly).

The foramen is located between the margins of the temporal bone (petrosal part) and the occipital bone (condylar part), and it comprises of three portions: two venous and one neural compartment.12 The venous compartments consist of a larger posterolateral venous channel, known as sigmoid portion and a smaller anteromedial venous channel, known as petrosal portion. The sigmoid portion receives blood flow from the sigmoid sinus and the petrosal portion from the inferior petrosal sinus. The petrosal portion empties into the sigmoid part through an opening at the medial wall of the jugular bulb, located between the IX cranial nerve anteriorly and the X-XI cranial nerves posteriorly.

Surrounding the dome of the jugular bulb in the temporal bone are numerous structures, including the posterior semicircular canal, the middle ear, the medial portion of the external auditory canal and the VII cranial nerve. The neural structures comprise the IX, X, and XI cranial nerves, located between the two venous compartments. The IX cranial nerve travels alone, slightly superiorly to the X and XI cranial nerves which are intimately associated with each other (Figs 4 to 6). The respective nerves penetrate the dura in the anteromedial margin of the intrajugular process, a fibrous or osseous bridge located between the temporal and occipital bones. Other structures related with the jugular foramen are: meningeal branches of the ascending pharyngeal and occipital arteries, cochlear aqueduct, Jacobson’s, and Arnold’s nerves.3 The glossopharyngeal nerve exits medially and superiorly off the jugular foramen, passing lateral to the internal carotid artery (ICA). The spinal accessory nerve leaves the jugular foramen medially to the jugular vein, usually passing anteriorly to the jugular vein extracranially, before going under the sternocleidomastoid muscle. The vagus nerve in found between the ICA and the jugular vein and is part of the vascular-nervous cervical complex. The hypoglossal nerve leaves the hypoglossal canal, passing behind the vagus nerve before coursing anteriorly off the ICA towards the tongue.
Glasscock-Jackson and Fisch classifications (Tables 2 and 3) have been used to classify the extension of glomus tumors. According to its extension and tumors, different approaches have been performed. The senior author considers the combined craniocervical approach the best way to achieve complete resection of glomus tumors. The control of the vascular structures involved and the wide visualization of the area are important for results. Although variations must be performed in accordance to each tumor peculiarities.

Tumors involving the jugular foramen are considered a challenge for the neurosurgeon due to the variable relationship among the neurovascular structures (jugular vein, carotid artery and cranial nerves) crossing the foramen through the skull base. The surgeon must be familiar with the origin site, growth pattern and geometry of each tumor.

**Table 2: Glasscock-Jackson classification**

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
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<tr>
<td>I</td>
<td>Small tumor involving jugular bulb, middle ear and mastoid</td>
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<tr>
<td>II</td>
<td>Tumor extending under internal auditory canal; may have intracranial extension</td>
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<tr>
<td>III</td>
<td>Tumor extending into petrous apex; may have intracranial extension</td>
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<tr>
<td>IV</td>
<td>Tumor extending beyond petrous apex into clivus or infratemporal fossa; may have intracranial extension</td>
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**Table 3: Fisch classification**

<table>
<thead>
<tr>
<th>Fisch Classification</th>
<th>Description</th>
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<tr>
<td>A</td>
<td>Tumor limited to the middle ear cleft (glomus tympanicum)</td>
</tr>
<tr>
<td>B</td>
<td>Tumor limited to the tympanomastoid area with no infralabyrinthine compartment involvement</td>
</tr>
<tr>
<td>C</td>
<td>Tumor involving the infralabyrinthine compartment of the temporal bone and extending into the petrous apex</td>
</tr>
<tr>
<td>C1</td>
<td>Tumor with limited involvement of the vertical portion of the carotid canal</td>
</tr>
<tr>
<td>C2</td>
<td>Tumor invading the vertical portion of the carotid canal</td>
</tr>
<tr>
<td>C3</td>
<td>Tumor invasion of the horizontal portion of the carotid canal</td>
</tr>
<tr>
<td>C4</td>
<td>Tumor invasion of the lacerum foramen and cavernous sinus</td>
</tr>
<tr>
<td>D1</td>
<td>Tumor with an intracranial extension less than 2 cm in diameter</td>
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<tr>
<td>D2</td>
<td>Tumor with an intracranial extension greater than 2 cm in diameter</td>
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I: Intradural extension, E: Extradural extension  
A = Glomus tympanicum; B, C and D = Glomus hypotympanicum
lesion affecting this respective region. It is a real advantage when undertaking a function-sparing procedure.

Tumors that arise within or penetrate in the jugular foramen lateral to the neural plane displace the nerves medially, a position favorable for their preservation during tumor extirpation. By contrast, medially positioned tumors displace the cranial nerves to the lateral tumor surface, presenting to the surgeon an unfavorable tumor location. Glomus tumors consistently arise at the lateral aspect of the jugular foramen, displacing the lower cranial nerves medially. This position accounts for the high rate of neural preservation in small and medium-size glomus tumors that have not invaded the foramen’s central partition. Meningiomas that arise laterally to the foramen (e.g., the posterior petrous surface, sigmoid sinus) favorably displace the lower cranial nerves medially. By contrast, tumors originating medially to the jugular foramen (e.g., clivus, foramen magnum) are unfavorable, displacing laterally the multiple small rootlets that coalesce the IX-XI cranial nerves into a vulnerable location. Schwannomas arise within the neural plane and have a variable geometry that depends partially on the nerve of origin. Theoretically, tumors that arise from the IX cranial nerve, located at the lateral surface of the neural plane, should be more favorable than those originating from the X or XI cranial nerves, located on its deep surface.

**Management of Glomus Tumors**

A complete examination of the cranial nerves is important to evaluate previous deficits and to define surgical routes. Audiometric studies are important to evaluate middle and internal ear function and should be performed in all cases. CT images are important for determining the extent of osseous involvement. Magnetic resonance imaging (MRI), with and without gadolinium enhancement, provides an excellent definition of the tumor and adjacent structures. Cerebral angiography consists in a useful tool for evaluating tumor’s blood supply and to define doubtable images caused by flow in MRI studies. Angiography may provide additional information about venous sinuses anatomy and flow aspects, involvement of the internal carotid artery (ICA), association between glomus and carotid body tumors, and torcular left-right drainage communication. Balloon occlusion test must be performed in some cases when ICA sacrifice is a possibility (Table 4).

**Embolization**

The use of preoperative embolization has been described as a great advance in the successful surgical management of glomus tumors. Embolization may reduce blood loss, the need of blood transfusion and may also shorten operative time. Despite these descriptions, the senior author does not use this method in all cases.

The preoperative arterial embolization is rarely complete in paraganglia tumors of the petrous bone. Intraoperative fluorescence angiography with indocyanine green (ICG) is a reliable procedure to evaluate the efficiency of preoperative embolization and may help the surgeon to estimate intraoperative bleeding. The identification of tumor blood supply branches represents a very important step in order to determine surgical strategy intervention. Ascending pharyngeal and stylomastoid arteries send branches to the respective tumors, consisting of the two most important vessels to be considered. The identification of such arteries and the transitory intrasurgical closing of the external carotid artery (ECA) correspond to the gold surgical management. Such a method avoids the high cost of the embolization. The senior author considers that one may reserve the preoperative embolization for D2 or D3 tumors of the Fisch classification or in those cases presenting lower cranial nerve deficits. The embolization should be performed in a period not superior to 72 hours previous to surgical treatment, because new afferent vessels might have blood flow, mainly by tympanic branches of the ICA. The senior author found that embolization may cause nerve dysfunction due to the density increase of the glomus tumors, making the resection an arduous process. Sometimes, the partial embolization may present worse results than not having embolization at all, once the tumor is modified and still highly vascularized.

**Radiation**

There are controversies about the role of the radiation therapy in the management of the glomus tumors. No controlled trials have been performed comparing surgery and radiation therapy. Some authors recommend its application on ancient patients or in those middle-age patients presenting important comorbidities. Some formal indications have been on residual or recidivating tumors. Glomus tumor rarely decreases in size following radiation therapy and the lack of growth is considered to indicate success. Radiosurgery and stereotactic radiosurgery consist of recent methods and may avoid complications whenever compared to conventional radiation therapy.

**Table 4: Evolution of management of glomus tumors**

<table>
<thead>
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<th>Evolution of the management of the glomus tumors</th>
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<tr>
<td>1950s</td>
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<td>1960s</td>
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<td>1970s</td>
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Stereotactic radiosurgery consists in an effective and safe treatment modality in the management of glomus jugulare tumors, particularly for residual or previously untreated small tumors. Studies show that radiosurgery results are similar in glomus jugulare tumor control and a superior morbidity profile as compared to surgical treatment alone. In addition, patients treated with radiosurgery usually remain clinically stable or present improvements. Given the indolent nature of these tumors, some groups consider radiosurgery, the first-line treatment of glomus lesions because of low morbidity and stabilization of the tumor growing. Embolization followed by radiosurgery may be an effective way for controlling pulsatile tinnitus produced by glomus tumors, probably because of the double effect of such therapy in the tumor blood supply. However, histological studies based on previously irradiated glomus tumors conclude that the tumor cells appear unaffected by irradiation. Catecholamines secretion is usually not affected by radiation therapy. However, a recent publication has described a case presenting secretion rate reduction in irradiated glomus tumors. This would be a significant finding. However, the patient may not be cured by this method. The resection-recurrence rate is affected when tumors are previously irradiated. In this cases the morbidity of the surgery is increased because of irradiated field. Besides those arguments, radiation therapy has not been shown to increase cranial injury and cerebrospinal fluid (CSF) leaking, presenting a great argument against surgery. Broader studies and follow-up must be developed in order to ensure the real benefits and complications related to the technique.

**Surgery**

The role of surgical procedures treating glomus tumors has proved its excellence through the decades. However, the surgical management of this challenging pathology is associated with severe injuries (Fig. 7). Surgery has been related with variable morbidity rates, but it is an effective modality treatment and the unique method that may immediately eradicate the tumor. The proportion of the resections has been increased along the years. Microsurgical techniques, advances in anatomy of the neural regions, the better understanding of adjacent structures and the performance of a good neuroanesthesia have contributed widely to achieve complete resections with low-rate complications. Cranial nerve palsies consist in possible complication and some authors have associated the invasiveness and clinical presentation comparing to postoperative results. Complete excision rates are variable and directly related to tumor size and its extension. In the senior author’s series, complete tumor extirpation was achieved in 91% of the cases. Between the period from December 1997 through December 2007, 34 patients (22 females and 12 males) with glomus jugulare tumors were treated. The mean age was 48 years and the mean follow-up was 52.5 months. Clinical findings included hearing loss in 88%, swallowing disturbance in 50%, and facial nerve palsy in 41% cases. MRI demonstrated a mass in the jugular foramen in 100% of the cases, a mass in the middle ear in 97%, a cervical mass in 85% and an intradural mass in 41% of the cases. The tumor was supplied by the ECA in all cases, by the ICA in 44%, and by the vertebral artery in 32% of the cases. Preoperative embolization was performed in 15 cases (44%).

### ANESTHETIC CONSIDERATIONS

Whenever dealing with glomus tumors, it is important to know those kinds of tumors that may produce and
segregate vasoactive amines. Due to such a condition, the anesthesiologist must be aware and be prepared to treat critical blood pressure variations that may occur. An anti-edematous therapy is also important to be considered. Once when working at the surface of the venous sinuses, the position of the patient and anesthetic management is mandatory to reduce surgical time and facilitate the surgeon’s work. Drilling above tense/congestive venous structures may be difficult and dangerous. Muscular relaxants can be used only during the beginning of the procedure, because facial nerve monitoring (Fig. 8) is not useful if the patient is under curare relaxation.

**SURGICAL TECHNIQUE**

**Position**

The senior author utilizes a supine position with the head fixed in a 3-pin head holder and rotated 45° contralateral to the lesion (Fig. 9). The homolateral shoulder is extended and posteriorly displaced. The abdominal region and the anterolateral region of the leg are prepared to resect fat and fascia if necessary. The incision consists in an arciform fashion, presenting a superior limit located 2 cm superior of the auricular conchae, a posterior limit 2 cm posterior to the mastoid process and an inferior limit placed 3 cm inferiorly from the apex of the mastoid process. One must be aware about the compression of the contralateral venous structures by excessive tilt of the head, once it may reduce venous drainage, making the surgical procedure difficult due to the increase of bleeding.

**Surgical Steps**

**Cranial Region Dissection**

- Arciform craniocervical incision
- Anterior retraction of the subcutaneous tissue with hooks, identifying the anterior margin of the sternocleidomastoid muscle, posterior margins of the external acoustic meatus and the posterior portion of the temporal muscle
- On the external surface of the sternocleidomastoid muscle, the auricular magnus nerve and the external jugular vein are identified. Auricular magnus preservation is important in cases that neural anastomosis needs to be performed (Figs 10A and B).
- The temporal muscle fascia (TMF) is dissected from the muscular fibers of the temporal muscle (TM) and displaced posteriorly and inferiorly, continuing with the insertion of the sternocleidomastoid muscle and splenius capitis muscles, exposing the mastoid apex and suboccipital region.
- Dissection of the posterior portion of the temporal muscle from the squamous portion of the temporal bone and is displaced anteriorly, exposing the

![Fig. 8: Electrodes to perform facial monitoring (Neuroalert®)](image1)

![Fig. 9: Surgical position to operate on jugular glomus tumors](image2)
posterior part of the zygomatic arch. The posterior part of the digastric muscle (DM) is dissected and separated anteriorly.

Cervical Dissection
- Identification of the common carotid artery (CCA) and its bifurcation into external carotid artery (ECA) and internal carotid artery (ICA)
- Identification of the internal jugular vein (IJV)
- Cranial nerves X, XI and XII are identified and preserved
- External carotid artery, as well as occipital and ascending pharyngeal arteries, are coagulated, ligated or sectioned
- The stylomastoid process is maintained intact to protect the extratemporal segment of the facial nerve
- Mastoidectomy, with visualization of the semicircular channels, sigmoid sinus, sphenoidal angle and middle fossa base (Figs 11 and 12). Abundant and permanent irrigation consists in an important procedure in this step. The protection of the facial nerve and dissection of the sigmoid sinus depend on this step.

Five Crucial Points in the Management of Glomus Tumors
1. Early vascular control (ICA and ECA)
2. Identification and coagulation/ligation of the afferent vessels
3. Proximal control of the sigmoid sinus and IJV
4. Identification and preservation of the lower cranial nerves and facial nerve
5. Obliteration and reconstruction of the surgical cavity.

In the last decades, many approaches have been developed to access glomus tumor of the jugular foramen. Basically, they vary in the amount of the temporal bone resection and dissection of the soft tissues. Independent of the approach discussed, the management of the facial nerve is considered the most important point during the procedure. In 1952, Capps described the anterior facial transposition technique, until Fisch and Pillsbury (1979) ruled the technique of the definitive anterior transposition, describing the necessity of the fixation of the nerve
Farrior (1984), Maniglia et al. (1992) and Martin and Prades (1992) have described the preservation of the facial nerve in situ; in its origin and inside the fallopian canal. Pensak and Jackler (1997), developed the technique called “Fallopian Bridge” that maintain the facial nerve in its canal and proceed with the resection of the tumor in an anterior, inferior and posterior way. 

Facial nerve manipulation has been associated with severe conmorbidity. Advising about it, the senior author suggests working anteriorly (prefacial), inferiorly (infrafacial) and posteriorly (retrofacial) to the facial nerve, maintaining the nerve in the canal most of cases. Is evident that in giant tumors or more difficult cases, rerouting of the nerve or including anastomosis with or without nerve grafts should be considered. The anastomosis with XII cranial nerve with its variants can be used as an option, especially in cases of neural invasion. Despite its reconstruction, facial nerve function is not as expected in most of cases.

The relationship between the facial management and the extension of tumor resection has been the main point to justify the anterior transposition of the nerve. In 1982, Fisch performed facial nerve anterior transposition in all cases of the glomus jugular tumors treated, presenting its functional recovery grades 1 or 2 (House and Brackman) in 87% and total tumor resection in 82% of all cases. Senior author found in 91% of total resection without necessity of facial transposition and described different approaches to manage glomus tumors. The type A consists in the infralabyrinthine retrofacial approach. Similar technique has been utilized by Maniglia and Martin and Prades while dealing with small or non-glomus tumors. The tumor in this zone is removed by aspiration, coagulation, and impaction of the venous canals that enter the jugular bulb, located in the inferior petrosal sinus and the internal condylar vein. In the author’s series, this respective approach has been performed in 11 patients, presenting one of them recidivating a tumor, despite the fact that postoperative magnetic resonance imaging (MRI) showed total resection. This approach is indicated in cases the tumor is not vascularized by the tympanic branches of the ICA or the anterior extension of the tumor does not reach the ICA. The technique has the advantage of the preservation of the facial nerve anatomy and function associated to the integrity of the vestibulocochlear structures and external acoustic meatus permeability. The facial preservation was achieved in all cases of the series, presenting one patient a transient facial palsy that recovered completely.

Fig. 12: View of the left craniocervical laboratory dissection. A mastoidectomy was done to identify the sigmoid sinus, the jugular bulb, facial nerve and semicircular canals in the parotid gland. When approaching glomus tumors, the decision about the anterior transposition of the facial nerve preconized by Fisch and Pillsbury ends up on discussion. A posterior transposition technique also has been described by House and Hitselberger (1976), but the devascularization of the facial nerve may result in ischemia and postoperative deficit. The respective technique was practically abandoned because of the high risk of postoperative transient or permanent alteration of the facial nerve. The amount of the tumoral resection that can be achieved was not so good as with anterior transposition.
The “fallopian bridge” technique was utilized by the senior author in 21 patients; middle ear structures were preserved in 7 patients and in 14 of them the respective structures were removed and the external acoustic meatus occluded. The risk of preserving in some cases structures of the middle ear is related with higher risk of fistula, that can occur through Eustachian tube or external auditory meatus. The disadvantage of this occlusion consists in the high rate of cholesteatomas formation and infection. In these cases, it is fundamental to complete drilling the mastoid cells, to effect resection of the tympanic membrane, and mucosa in the promontorium area. Senior author advises the only situation that indicates meatus occlusion and middle ear structures removing when tumor grows cranially to the petrous portion of the ICA. Facial nerve anastomosis has been performed in two cases of the senior author series. The technique chosen was initially preconized by Hammerschlag (1999), consisting of nervous graft terminolateral suture to the hypoglossal nerve and terminoterminal in the facial nerve. Functional recovery was grade 4 of House and Brackman classification with light hemi tongue atrophy.

The management of the glomus tumors are based on the following criteria:

- The blood supply of the tumor
- Functional state of the facial nerve
- Extension of the tumor.

The approach may be patterned as follows:

A. Infralabyrinthine: Indicated to resection of tumors receiving the main blood supply from the ECA, without anterior extension or around the ICA in its ascending segment (Figs 13A to D).

B. Fallopian bridge: Tumor visualization anteriorly and posteriorly to the facial nerve, without occlusion of the external acoustic meatus or resection of middle ear structures. It is indicated when tumor involves the ICA in the ascending way and tympanic branches of the ICA blood supply contribution (Figs 14A to C).

C. Similar to B, with occlusion of the external acoustic meatus and resection of middle ear structures. It is reserved to those tumors presenting cranial extension, following the ascending pathway of the ICA through the cavernous sinus (Figs 15A to C).

D. Anterior extension: Reserved to those tumors presenting wide anterior extension involving the ICA in its complete petrous segment, with extension to cavernous sinus, and also when the patient presents clinical symptoms and signs of hearing and facial dysfunction (Figs 16A to C).

As discussed previously, cranial nerves preservation consists in another important aspect of the surgical management. Anatomically, the nerves IX, X and XI are in relation with the medial wall of the Jugular vein and Jugular bulb. Considering this information, the medial portion of this vein is a crucial area for preservation of the neural structures (Figs 17A to C). Called by Al-Mefty and Teixeira in 2002 as “intrabulbar resection”, when operating glomus tumors, it is important to preserve a portion of the jugular bulb and the wall of the jugular vein in order to avoid damaging nerves. According to Lusting and Jackler (1996), CT and MRI (Magnetic Resonance Imaging) are very helpful whenever showing a regular middle bone surface as a great chance that nerves may not be invaded by the tumor. In contrast, intracranial extension has been related with postoperative lesion of nerves and non-complete tumor resection (Jackson et al. 1991).

**Types A, B, C and D Approaches**

The approach was tailored to each patient and classified in four types of them (Table 5). The infralabyrinthine retrofacial approach (Type A) was used in 32.5%; infralabyrinthine pre and retrofacial approach without occlusion of the external acoustic meatus (Type B) in 20.5%; infralabyrinthine pre and retrofacial approach with occlusion of the external acoustic meatus (Type C) in 41%; and the infralabyrinthine approach with transposition of the facial nerve and removal of the middle-ear structures (Type D) in 6% of the patients. Radical removal was achieved in 91% of the cases and partial removal in 9%. Among 20 patients without preoperative facial nerve dysfunction, the nerve kept its anatomical position in 19 (95%), and facial nerve function was normal during the immediate postoperative period in 17 (85%). Six patients (17.6%) had a new lower cranial nerve deficit; recovering of swallowing function was adequate in all cases. Voice disturbance remained in all six cases. Cerebrospinal fluid leakage occurred in six patients (17.6%), without the need of reintervention. One patient died in the postoperative period due to pulmonary complications. The global recovery, based on the Karnofsky Performance Scale (KPS), was 100% in 15% of the patients, 90% in 45%, 80% in 33% and 70% in 6%. Considering the experience of the senior author, radical
Figs 13A to D: Magnetic resonance imaging (MRI). (A) Sagittal contrast-enhancing sequence; (B) Axial contrast-enhancing sequence showing a right glomus tumor approach by infralabyrinthine retrofacial (Type A Approach); (C) Angiography showing the vascularization of the tumor from branches of the external carotid artery; (D) Postoperative MRI study in axial sequence that shows complete resection of the lesion (TU: Tumor)
Figs 14A to C: (A) Magnetic resonance imaging (MRI) axial sequence with contrast enhancement showing a left jugular glomus tumor. Type B approach has been performed; (B) Angiography with blush in the region of the tumor; (C) Axial MRI sequence that shows complete resection of the tumor (TU: Tumor)
Figs 15A to C: (A) Magnetic resonance imaging (MRI) axial sequence with contrast enhancement; (B) Angiography that shows a large left glomus tumor; (C) Postoperative MRI. The tumor was completely removed (TU: Tumor)
Figs 16A to C: (A) A very large tumor demonstrated by this MRI with contrast enhancement in axial sequence; (B) Angiography that shows the extended tumor highly vascularized, with participation of the internal and external carotid artery branches; (C) Postoperative MRI in axial sequence with contrast enhancement. The tumor completely resected.
removal of glomus jugulare tumor may be achieved without anterior transposition of the facial nerve. However, the extension of dissection should be tailored to each case based on tumor blood supply, preoperative symptoms, and tumor extension.

CONCLUSION

Glomus tumors are challenging craniocervical lesions. Surgical treatment is the only procedure that can immediately cure the patient. Anatomical knowledge and practice are crucial items to minimize surgical complications. The operative field provided by the retrofacial infralabyrinthine approach, or the pre and retrofacial approaches, with or without closure of the external acoustic meatus, allows a wide exposure of the jugular foramen area. Global functional recovery based on the KPS was acceptable in 94% of the patients. Despite surgical management as described by the senior author, one should be aware of the variety of management options and its results. Treatment should be based on the age, clinical condition, tumor localization and its extension.

GLOMUS JUGULAR TIPS

1. Glomus tumors are rare tumors and consist in the most frequent tumors located in the jugular region.
2. Females may be six times more affected than males.
3. Glomus tumors are highly vascularized tumors and the correct management must consider the vascular blood supply.
4. A complete examination of facial and lower cranial nerves must be performed. This aspect may help to define the best way to approach the lesion.
5. The most common clinical presentation consists in hearing disturbance, swallowing deficit and facial nerve palsy.
6. Facial mobilization should be avoided in most of cases. Working in infralabyrinthine, retro and prefrontal areas, we can achieve complete tumor resections.

7. Surgical treatment is possible in most of the cases, presenting low morbidity and excellent functional results.

8. The anatomical complexity of the area, the high vascularization of the lesion and the inadequate planning are the main causes of treatment failure.

9. Radiation therapy is effective and considering the benignity of the lesion, this modality should be considered as an option.

10. Tumor embolization must be considered in some cases.

11. Cerebrospinal fluid leakage consists in the most frequent complication of the surgical procedure, followed by lower cranial nerves dysfunction.

12. Fat, fascia and pediculated muscle rotation, reinforced by biological glues are the most important way to prevent fistula.

13. Like other cranial base approaches, you must program the close reconstruction before your opening approach.

14. Remember that variants like anomalous venous drainage, high jugular bulbs or dominants sinus can mimic tumors according to the flow pattern. Complete clinical examination and studies with CT, MRI and arterial/venous angiography can help you to establish the diagnosis.

REFERENCES


