ORIGINAL ARTICLE

Surgery of Glomus Jugulare Tumors

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ABSTRACT

The treatment of choice for glomus jugulare tumors is still controversial. High rates of morbidity, incomplete resection, and the aggressive behavior of these tumors are the main arguments for advocates of primary radiotherapy. However, constant refinements in skull base techniques have made complete resection of these lesions a realistic goal. The high probability of achieving local control of these tumors by surgery has convinced us to support this option strongly. Between 1993 and 2000 we diagnosed 52 glomus tumors of the temporal bone. Of these patients, only 42 had a class C lesion (glomus jugulare) and were included in this study; 37 of these patients underwent surgery, 10 of whom had intracranial extension of the disease. The overall resection rate was 96%. Facial nerve function at 1 year was House-Brackmann grade I to II in 52% of patients and grade III or better in 84% of patients. Hospitalization was shorter than 14 days in 33 patients (89%). All patients with pharyngolaryngeal palsy had sufficient compensation at discharge. Twelve vocal chord Teflon injections were performed after surgery to reduce hoarseness and aspiration. No patient died. No relapse was observed (mean follow-up, 4.9 years).

KEYWORDS: Glomus jugulare tumor, infratemporal approach, pharyngolaryngeal paralysis

The effectiveness of surgery compared with radiotherapy as the primary treatment for glomus jugulare tumors remains controversial. A definitive consensus on the best treatment modality for this disease cannot be reached because there are few studies and most of them lack adequate information. Much of the criticism against the surgical management of glomus jugulare tumors is based on the morbidity rates associated with cranial nerve loss and the common belief that radical removal is almost impossible because these tumors are histologically aggressive. This study evaluated the role of surgery

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in the management of glomus jugulare tumors by reviewing our results with long-term sequelae and radical resection.

MATERIALS AND METHODS

From January 1993 to October 2000, 52 patients were diagnosed as having glomus tumors of the temporal bone. Of this group, 42 patients (28 females, 19 males) had glomus jugulare tumors (class C tumors). The other 10 patients, diagnosed with glomus tympanicum tumors (class A and B tumors), were excluded from this series.

Tumors were classified using the Fisch classification (Table 1).[1] Of the 42 patients with a class C tumor, 3 had previously undergone surgery. None had undergone external beam radiation, and 39 patients had had no previous treatment. Eight patients (20%) had multicentric, synchronous lesions, 4 of whom had familial distribution of the disease. The most common combinations were with carotid body (5) and glomus vagale (3) tumors. Three patients had bilateral temporal lesions.

On otoscopy 33 patients (80%) with glomus jugulare tumors had a typical red middle ear or external auditory mass. The remaining patients had an infiltrating hypervascularized temporal bone mass documented by an anomalous injection of the vessels of the posteroinferior auditory canal. As with glomus tympanicum tumors, the most frequent otologic symptoms were pulsatile tinnitus, followed by hearing loss, mucopurulent or hemorrhagic otorrhea, otalgia, and vertigo (Table 2).

Twenty-nine patients (70%) had no preoperative cranial nerve deficit. Deficits were most frequently associated with the eighth cranial nerve, followed by the tenth, the ninth, the twelfth, the seventh, and the eleventh (Table 3). Hoarseness and dysphagia were present, respectively, in 5 (12.5%) and 2 (5%) patients, and facial weakness in 5 (12.5%). Preoperative facial nerve function was normal in 37 patients (88%) while 5 patients (12%) exhibited facial weakness. One patient (2.5%) had a catecholamine-secreting tumor.

All patients underwent a detailed neuroradiological evaluation with high-resolution computed tomography, gadolinium-enhanced magnetic resonance imaging (MRI), and conventional or selective digital subtraction angiography.

Table 1 Fisch Classification of 52 Glomus Tumors of the Temporal Bone

Type of Tumor	No. Patients
Glomus Tympanicum	
Class A	4
Class B	6
Glomus Jugulare Tumors Class C	
C1	16
C2	17
C3	6
C4	3
Intracranial Extension	
Class D	12

Table 2 Otologic Symptoms Associated with 42 Glomus Jugulare Tumors

Symptoms	No. Patients (%)
Pulsatile tinnitus	36 (86)
Hearing loss	29 (69)
Otalgia	8 (19)
Otorrhea	3 (7)
Vertigo	9 (21)

Table 3 Preoperative Cranial Nerve Deficits Associated with 42 Glomus Jugulare Tumors

Cranial Nerve	No. Patients (%)
V	0 (0)
VI	1 (2.5)
VII	5 (12)
VIII	9 (22.5)
IX	7 (17.5)
X	8 (2)
XI	6 (15)
XII	3 (7.5)
None	29 (70)

Permanent balloon-occlusion of the internal carotid artery (ICA) was performed in 5 patients with C3 to C4 glomus tumors when angiography documented an intimate relationship with the horizontal segment of the ICA and cavernous sinus and a significant blood supply from petrous or cavernous branches of the ICA. Our protocol for ICA occlusion involves angiography with cross compression and a temporary intraluminal balloon-occlusion test. In 1 patient both the carotid and vertebral artery had been occluded for direct involvement of the two arteries. None of the 5 patients had any immediate or delayed neurological deficits.

One patient did not tolerate occlusion of the ICA, and the balloon was immediately deflated with no permanent neurological consequences. Positron-emission tomography scanning of the head was positive in 3 high-risk patients even though they passed the balloon occlusion test. In 8 patients with C2 and C3 tumors, simple functional angiography was performed to assess the competence of the circle of Willis. A balloon occlusion test was performed in 3 of these patients. All 37 surgical patients underwent preoperative embolization with polyvinyl alcohol microparticles, lyophilized dura, or histoacrylic glue.

Exclusive external beam radiation was used in 2 older patients and in 1 patient who refused surgery. No treatment was recommended for 2 patients who were older than 80 years. Eight D1 intracranial extensions were removed in a single-stage operation. Of the 4 patients with a D2 intracranial extension, 2 underwent a second stage through a petro-occipital trans-sigmoid approach. The follow-up included MRI 1 year after surgery and every 2 years thereafter.

Surgical Technique

Conservative jugulopetrosectomy and infratemporal fossa approaches represent different surgical options for the management of glomus jugulare tumors. The latter was used in 33 patients (Table 4). A conservative jugulopetrosectomy that preserved the facial nerve was performed in 4 patients.

Table 4 Surgical Approaches to 37 Glomus Jugulare Tumors

Approach	No. Patients
Conservative jugulopetrosectomy	4
Infratemporal fossa type A	28
Infratemporal fossa types A and B	5
Petro-occipital Trans-sigmoid (2nd stage)	2

Conservative Jugulopetrosectomy

This approach preserves the normal anatomy of the external and middle ear. It is indicated for small tumors confined to the jugular foramen and infralabyrinthine area with minimal involvement of the ICA at its foramen.[2] An incision placed about 6 cm behind the postauricular crease and extended inferiorly and superiorly exposes the neck and temporal bone. The seventh, ninth, tenth, eleventh, and twelfth cranial nerves are followed to the skull base. The seventh cranial nerve is dissected until its bifurcation. The major vessels of the neck are identified. The internal jugular vein is ligated and divided to obtain distal control of the vein. A complete enlarged mastoidectomy is performed, and the posterior wall of the bony ear canal is preserved. The lateral sinus is exposed from the transverse sinus to the jugular bulb and then opened and packed intraluminally with Surgicel to obtain proximal control of the lateral sinus. The facial nerve is identified and skeletonized to the stylomastoid foramen. An extended facial recess is drilled to enable the initial visualization of the tumor. When the external auditory canal is preserved, the nerve is usually mobilized from the second genu laterally to expose the jugular bulb. Further inferior widening of the extended facial recess and removal of the tympanic bone anteroinferiorly expose the lower part of the tympanic segment of the ICA (infratubal portion) and the carotid fora-

men. Once the ICA is freed from the tumor, it is mobilized from the infralabyrinthine area. With the internal jugular vein and the lateral sinus identified, the tumor is resected from the skull base and cranial nerves. If possible, the medial wall of the jugular bulb is left intact to avoid danger to the cranial nerves at the pars nervosa of the jugular foramen.

Infratemporal Fossa Approach

When the tumor extends beyond the jugular foramen anteriorly in the petrous bone and distal exposure of the petrous carotid artery is needed, the surgical procedure must be expanded. This approach sacrifices the external and middle ear, but it offers access to the infralabyrinthine area, petrous apex, and, through the infratemporal fossa, to the entire petrous carotid artery, clivus, and cavernous sinus. The petro-occipital skull base, neck, and infratemporal fossa are accessed through a C-shaped incision that allows an anteriorly based flap. The external ear canal is then transected and sutured. Initial cervical exposure proceeds as it does for smaller tumors. Both temporozygomatic and cervicofacial branches of the facial nerve are exposed to allow sufficient mobilization of the nerve from the parotid for anterosuperior transposition. The skin of the external auditory canal, the tympanic membrane, and the ossicles are removed, and a radical mastoidectomy is performed. The lateral sinus, middle fossa dura, and facial nerve are identified.

The lateral sinus is unroofed. After the internal jugular vein is ligated in the neck, the vessel is opened and packed intraluminally as described. Exposure of the petrous ICA requires anterior dislocation of the ascending ramus of the mandible, which, in turn, requires anterosuperior rerouting of the facial nerve. This delicate maneuver is mandatory to obtain satisfactory control of the jugular foramen and petrous carotid artery. Once the facial nerve has been mobilized from the geniculate ganglion laterally, anterior reflection of the mandible, maintained by the Fisch infratemporal fossa retractor, allows exposure of the glenoid and posterior infratemporal fossae. The removal of the medial wall of the glenoid fossa and of the bony eustachian tube exposes the vertical portion of the petrous carotid artery and of the anterior pole of the tumor. Subtotal petrosectomy permits control of the superior pole of the tumor and its radical dissection from the cancellous bone of the petrous apex and from the petrous carotid wall (infratemporal approach type A).

Further tumor extensions involving the petrous apex, foramen lacerum, cavernous sinus, and clivus need extended approaches. Removal of extensions requires switching from the infratemporal type A to the type B or C approach. This change creates some problems with the facial nerve, which must be transposed from anteriorly to inferiorly to expose the infratemporal fossa.[3] Temporary resection of the posterior portion of the zygomatic arch and inferior reflection of the temporal muscle and of the mandibular condyle with the Fisch retractor opens the entire infratemporal fossa and its anatomy. The middle fossa dura is skeletonized to expose the foramen spinosum and foramen ovale. The middle meningeal artery and mandibular division of the trigeminal nerve are coagulated and divided. The more medial eustachian tube must be resected to expose the horizontal portion of the carotid artery (infratemporal fossa approach type B). Drilling the bone of the pterygoid process (infratemporal approach type C) gives full exposure of the foramen lacerum and lateral wall of the cavernous sinus.[1] Perfect control of the horizontal portion of the ICA allows tumor to be resected from the floor of the middle fossa dura, petrous apex, clivus, and the cavernous sinus, which is rarely infiltrated. If these extended infratemporal approaches are combined with the transcochlear route, they provide full access to the lateral skull base and all the anteromedial and intracranial extensions of the disease to be removed.[4]

Intracranial extension of the tumor into the subarachnoid space is common and can be accurately predicted by MRI. It usually occurs in the posterior fossa directly through the dura or through the lower cranial nerves at the pars nervosa of the

jugular foramen.[5] Single-stage resection of transdural glomus tumors is complicated by problems of dural defect reconstruction and cerebrospinal fluid management, which are more complex and troublesome in these cases than in other skull base procedures. When intracranial extension is limited, the resulting dural defect is small. In these cases unstaged tumor resection should be considered safe. For more extensive degrees of intracranial spread, especially when transcochlear or translabyrinthine exposures are indicated, it is best to leave the intradural extension in place. Secondary intracranial tumor resection through the posterior fossa and its contents is seldom a problem because the devascularized tumor dissects easily from the brain.[6]

RESULTS

Neurologic sequelae involved the facial nerve and lower cranial nerves.

Facial Nerve

Of the 37 patients who underwent surgery, the facial nerve was left in the fallopian canal and preserved in 4 cases (10%), infiltrated by tumor and therefore deliberately transected in 6 cases (15%), accidentally transected after tumor removal in 2 (5%), and permanently transposed in 25 (70%). The overall rate of anatomical preservation of the facial nerve was 80% (29 patients). A sural nerve grafting was performed in 6 cases. In 2 patients with longstanding paralysis the facial nerve was not repaired. Facial nerve function measured at 1 year was as follows: grade I in 7 patients (18%), grade I or II in 19 patients (51%), and grade III or better in 31 patients (84%). See Table 5.

Cranial Nerves IX and X

The ninth and the tenth cranial nerves were anatomically preserved in 18 (48%) and 13 (35%) patients, respectively (Table 6).

The function of cranial nerves IX and X was lost preoperatively in about 20% of cases and was caused by surgery in 50%. Most patients reported some degree of persistent dysphagia, hoarseness, and aspiration, but none was disabled by these symptoms. No patients have reported recurrent pneumonia; 12 patients have undergone postoperative vocal cord injection with Teflon paste.

Total excision was possible in most patients. The rate of complete resection was 100% for 13 C1 and 16 C2 tumors, 80% for the 4 C3 and 2 C4 tumors, and 84% for 10 class D tumors. The overall resection rate was 96%.

None of the patients with subtotally resected tumors has undergone radiation therapy. None of the 37 glomus jugulare tumors has recurred after total resection (mean follow-up, 4.9 years). There were no cerebrospinal fluid leaks through the wound. In 3 patients with class D lesions a subcutaneous

Table 5 Postoperative Functional Outcome of Facial Nerve in 37 Patients

House-Brackmann Grade	Left in Place No. (%)	Transposed No. (%)	Cable Grafted No. (%)	Longstanding Paralysis No. (%)
I	4 (100)	3 (12)	0	0
II	0	12 (48)	0	0
III	0	8 (32)	4 (66)	0
IV	0	1 (4)	2 (33)	0
V	0	1 (4)	0	0
VI	0	0	0	2 (100)

Table 6 Preservation of Cranial Nerves as a Function of Size of 37 Tumors

Tumor	No. Cases	CN VII No. (%)	CN VIII No. (%)	CN IX No. (%)	CN X No. (%)
C1	13	12 (92)	11 (85)	10 (77)	11 (85)
C2	16	12 (75)	7 (44)	3 (20)	7 (45)
C3	5	3 (60)	0	0	0
C4	3	2 (70)	0	0	0
Total	37	29 (80)	18 (50)	13 (35)	18 (42)

CN, cranial nerve.

cerebrospinal fluid collection resolved after sterile cerebrospinal fluid aspiration and application of compression dressing. No wound required revision. There were no perioperative deaths. One patient developed a stroke 2 days after surgery related to ICA thrombosis and was permanently hemiplegic. Postoperatively, 2 patients with acute respiratory distress syndrome required a temporary tracheotomy. Three patients developed partial ischemia of the wound, pinna, or both.

DISCUSSION

Surgery of glomus jugulare tumors usually requires extensive dissection of the posterolateral skull base. These tumors tend to arise in the osteodural interface of the jugular foramen and extend inside the petrous bone, cranium, and cervical regions. Inside the petrous bone these tumors spread from the jugular foramen along routes of least resistance that usually involve the fallopian and carotid canals.[7] Effective management of the facial nerve and major vessels of the petroccipital skull base is indispensable to obtain a successful outcome after excision of a glomus jugulare tumor.

The facial nerve represents an anatomic obstacle to surgical exposure of the jugular foramen and intrapetrous carotid artery. The management options for the facial nerve include a simple exposure, partial or complete mobilization, and segmental resection. Tumor size and control of the distal ICA are the main factors that determine facial nerve management. For disease confined to the jugular foramen and in which the ICA is not involved, simple exposure of the seventh cranial nerve is sufficient. However, excision seldom proceeds without endangering the facial nerve. Typically, the nerve must be mobilized. When the external auditory canal can be preserved, partial mobilization from the second genu laterally provides sufficient exposure of the tumor and control of the ICA. Postoperative facial nerve function tends to be excellent.

For more extensive tumors, complete exposure of the vertical portion of the petrous ICA is mandatory. This goal can be achieved only by anterior dislocation of the mandible combined with complete anterosuperior rerouting of the facial nerve from the geniculate ganglion laterally.[2] Paralysis of the nerve usually follows, but recovery is satisfactory (House-Brackmann grade I to III). Postoperative facial paralysis is rare and usually has a late onset. Preoperative facial paralysis always indicates direct invasion of the nerve. In such cases segmental resection is necessary. Continuity of the facial nerve can be accomplished via direct end-to-end anastomosis or graft interposition with or without rerouting. A grade IV to II is a reasonable outcome to expect. Intraoperative monitoring improves the chance of preserving neural integrity and significantly reduces the postoperative rate of permanent facial nerve dysfunction.[8]

Complete dissection of the tumor from the petrous ICA is probably the most limiting factor for achieving radical removal of glomus jugulare tu-

mors. Surgical exposure of the ICA must guarantee its proximal and distal control as well as visualization of almost its entire circumference to enable mobilization and surgical maneuvers. The tumor is usually dissected from the ICA in a subperiosteal plane. When a subadventitial plane is needed, extrication of the vessel is far more dangerous and may be infeasible. Preoperative balloon occlusion of the ICA is performed whenever neuroradiological investigations indicate direct involvement of the carotid wall.

Cranial nerve preservation during the surgical resection of glomus jugulare tumors is the main factor in reducing the postoperative rate of morbidity and directly depends on the size of the tumor. The facial nerve was sacrificed in 22% of our cases (8 patients). However, 12.5% of the patients had a preoperative nerve deficit. Sural nerve interposition of the sacrificed nerve performed in 6 patients resulted in acceptable functional outcomes: 66% (4 patients) were House-Brackmann grade III and 33% (2 patients) were House-Brackmann grade IV. Permanent transposition of the nerve resulted in good long-term functional outcomes: 23 patients (92%) were House-Brackmann grade III or better, and 15 patients (60%) were House-Brackmann grade I or II (Table 5).

Facial nerve outcome (Table 5) was evaluated 15 months after surgery. Cranial nerves IX and X had to be sacrificed in 65% and 50% of cases, respectively, resulting in permanent hemipharyngolaryngeal paralysis. Dysphagia, hoarseness, and aspiration are often severe perioperatively. In such cases, a small nasogastric feeding tube was left in place 10 to 12 days to prevent pneumonia and to allow spontaneous compensation. When dysphagia persisted, a Teflon paste injection of the paralyzed vocal cord improved both voice and aspiration in all cases. No temporary or permanent gastrostomy was necessary. Most patients (89%) were sent home within 2 weeks of surgery. Cerebrospinal fluid leaks and meningitis were absent in our patients, even in the ones with class D lesions. This finding compares favorably with other series.[9,10] For large intradural tumors that involve the brain stem, even if free flaps have greatly reduced the risk of cerebrospinal fluid leaks, we still prefer a staged petro-occipital removal.[11]

CONCLUSIONS

In the past 10 years with the growing emphasis on quality of life, attention has drifted away from issues of tumor resectability, which is possible in most patients, toward functional outcomes. The consequences of surgical resection are predictable, and the neurotological deficits can often be rehabilitated. Surgical resection of jugular foramen lesions is most often criticized on the basis of the morbidity following lower cranial nerve loss. Cranial nerve preservation depends on tumor size. With small lesions where the rate of cranial nerve preservation exceeds 80%, functional outcomes are excellent. With larger lesions cranial nerve loss is often present preoperatively, and surgery does not always create new deficits. In our series 12.5% of the patients had preoperative facial palsy while 20% had lower cranial nerve deficits.

When acute cranial nerve losses must be created, they can be compensated for or efficiently rehabilitated, especially in young patients. The same cannot be stated for older patients (60 years or older), where the risks of aspiration and ab ingestis pneumonia strongly discourage surgery. A patient's age should be considered the main factor in reducing postoperative rates of morbidity and mortality related to pharyngolaryngeal paralysis. Surgery in patients older than 60 years with C2 to C3 lesions should be discouraged, even when a compensated cranial nerve deficit is present.

One year after surgery, facial nerve outcome was House-Brackmann grade III or better in 84% of our patients and grade I or II in 52%. Twelve pa-

tients were treated with vocal cord Teflon injection to reduce hoarseness. The other patients who suffered hemipharyngolaryngeal palsy had sufficient compensation and exhibited no evidence of aspiration at follow-up.

The relative efficacy of surgery compared to radiation therapy as the primary treatment for glomus jugulare tumors remains controversial. Literature supports both modalities, and the conclusions drawn in different reports have often been extreme. This inability to reach a definitive consensus on the issue probably reflects the lack of adequate information regarding patient selection, staging, and follow-up. Available data are from small series or from extremely heterogeneous series where tumor size, previous treatments, and complication rates are not clearly reported and do not allow results to be compared.[12–25]

In young patients, especially those with small lesions, neurotologic conditions make surgery less attractive. Nevertheless, we believe that surgical eradication of the tumor rather than alteration of its biological potential represents the safest treatment modality in these patients. In elderly patients with advanced disease, radiation must be preferred. Between these two extremes, there is a wide range of situations in which individualized approaches based on various parameters such as size of the tumor, age, existing nerve deficits, performance status, social conditions, motivation, and cooperation should be considered.

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