Transmastoid-Infralabyrinthine Tailored Surgery of Jugular Paragangliomas

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ABSTRACT

This article presents our experience with the management and outcomes of patients with sporadic glomus jugular tumors using a tailored surgical approach. We conducted a retrospective study of 49 patients with sporadic jugular paragangliomas. Thirty-eight patients (78%) were treated using the transmastoid-infralabyrinthine (TM-IL) approach, and 11 (22%) were treated with the infratemporal fossa type A (IFT-A) approach. Mean follow-up period was 8.2 years. Surgical cure was achieved in all but 6 patients using the TM-IL approach (84%) and in 8 of 11 patients with the IFT-A approach (73%). A House-Brackmann grade of I or II facial nerve function was obtained postoperatively in 84% of TM-IL cases and in 56% of the IFT-A cases. The incidence of other complications—meningitis and transient cerebrospinal fluid leak—was higher in the TM-IL group (8% and 13%, respectively) compared with the IFT-A group (0% and 9%, respectively). We concluded that the TM-IL approach with individually tailored management of the facial nerve fulfills the long-term requirements of both keeping good postoperative facial function and obtaining adequate tumor resection.

KEYWORDS: Jugular paraganglioma, surgery, facial nerve, transmastoid-infralabyrinthine approach, infratemporal fossa approach

The resectability of jugular paraganglioma (PGL) is influenced by a variety of factors including bone invasion, infiltration of lower cranial nerves, involvement of major blood vessels, and spread into the intradural space. The prerequisite for safe and complete tumor removal is adequate exposure. For this reason, surgical management of the intratemporal portion of the facial nerve (FN) is a challenge. A balance has to be struck between maintaining good postoperative facial function and complete or ad-

equate tumor resection. Anterior transposition of the FN from the geniculate ganglion to the stylomastoid foramen, as described by Fisch in 1977,¹ allows superior exposure of the tumor, including its anterior spread along and beyond the internal carotid artery (ICA). The preservation rate of normal postoperative FN function with this technique was reported to vary between 65.8 and 80%.^{2,3} Problems experienced by others using this technique resulted in alternative solutions to avoid anterior transposition.^{4–11} With

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these techniques, 70 to 95% of patients retained normal postoperative facial function.^{6,11–15}

However, the FN function is just one of the important aspects of jugular PGL surgery. The dilemma of whether partial FN transposition, which allows preservation of the external and middle ear structures, risks incomplete tumor removal and surgical complications (e.g., meningitis) is still unresolved. Because long-term data are insufficient, we have analyzed our data using two different surgical techniques to give insight on the likely outcomes following the transmastoid-infralabyrinthine approach (TM-IL) and the infratemporal fossa type A (IFT-A) approach. In this way, we have developed a tailored approach to these tumors to maximize FN preservation.^{16–18}

MATERIAL AND METHODS

Patient Population

Forty-nine patients with sporadic jugular PGLs were treated surgically by the first author (M.G.) during the years 1990 to 2003 at the ENT University departments in Erlangen and Frankfurt,

Germany, and Zagreb, Croatia. Of these, 39 were women and 10 were men. Their mean age at the time of surgery was 57 years (range, 24 to 73 years). None of the patients presented with malignant tumors or catecholamine-secreting tumors. The Fisch and Mattox system was used to classify the tumors by size and spread (Table 1).¹⁹ There were 20 C1 (41%), 24 C2 (49%), and 5 C3 (10%) tumors. Thirty-three tumors (67%) had intracranial extensions, of which 23 were extradural (47%) and 10 were intradural (20%).

Surgical Approach

Thirty-eight patients (78%) were treated using the TM-IL approach, and 11 patients (22%) underwent the IFT-A approach. In two patients, the TM-IL approach was combined with the enlarged middle fossa or the retrosigmoid approach to gain adequate exposure of the tumor.

Highlights of the TM-IL approach include the following steps. The patient is placed supine with the head turned to the opposite side. Intraoperative facial, glossopharyngeal, and vagal nerve monitoring is used routinely. A C-shaped, postauricular incision is made allowing exposure of

Table 1 Tumor Size, Surgical Approaches, and Radicalism of Surgical Resection

Tumor Size	No. of Patients	TM-IL ± MF/RS	Recurrence	IFT-A	Recurrence
C1	4	4	0	0	0
C1De1	5	4	0	1	0
C1De2	4	4	0	0	0
C1De2Di1	5	4	2	1	0
C1De2Di2	2	1	0	1	0
C2	5	5	0	0	0
C2De1	6	5	0	1	0
C2De2	6	4	1	2	0
C2De2Di1	5	4	2	1	0
C2De2Di2	2	0	0	2	2
C3De2	2	0	0	2	1
C3De2Di2	3	3	1	0	0
Total	49	38	6	11	3
			16%		27%

TM-IL, transmastoid-infralabyrinthine; MF, middle fossa; RS, retrosigmoid; IFT-A, infratemporal fossa type A.

the temporal bone, lateral skull base, and neck. The external auditory canal (EAC) is not transected. The neurovascular structures of the neck (internal jugular vein, ICA, external carotid artery and its branches, and the lower cranial nerves IX–XII) are exposed and secured with vascular slings. The ascending pharyngeal and occipital arteries are ligated. The FN is identified at the stylomastoid foramen. A complete mastoidectomy with exposure of the sigmoid sinus, labyrinth, and middle and posterior fossa dura is performed.

Further steps are tailored according to each patient's requirements. These are based on the extent and exposure of the tumor. The FN is identified at the second genu and in the inferior portion of the mastoid using the fascia of the digastric muscle as a landmark. The fallopian canal is skeletonized using a diamond burr (Fig. 1). A thin shell of bone is left over the nerve. Once the nerve is defined, bone medial to it is removed, including the retrofacial and anterofacial air cells and the hypotympanic bone. In this way, good exposure of the sigmoid sinus and jugular bulb is achieved. The skin of the ear canal and the tym-

panic membrane is elevated to expose the middle ear and to assess tumor extension and exposure. The mastoid tip is removed. At this stage, the lower segment of the vertical segment of the ICA is completely exposed. Tumor removal can then be accomplished anteriorly and posteriorly to the FN canal.

By superior reflection of the soft tissues of the external canal, together with adequate tilting of the operating table, the ICA can be exposed up to its genu at the level of the eustachian tube. The patient's head and the microscope are carefully positioned to allow a good view of the infralabyrinthine compartment of the temporal bone. This exposure allows control of the entire circumference of the ICA in its vertical course. Additional exposure of the artery, if required, can be achieved by rerouting the FN from the second genu only. In this way, the integrity of the external auditory canal can be preserved (Fig. 2). The middle ear component of the tumor is then delivered into the jugular fossa. Tumor involving the ICA is removed by establishing a plane of cleavage along its adventitia. By removing the infralabyrinthine air cells, it is possible to follow the tumor medially to the

Figure 1 The fallopian canal is skeletonized with a diamond burr from the second genu to the inferior portion of the mastoid. A thin shell of bone is left over the nerve. The facial recess is wide open, and the external ear canal kept intact.

Figure 2 The facial nerve is rerouted from the second genu keeping the soft tissues of the external auditory canal still preserved. The low posteroinferior visualization of the infralabyrinthine compartment allows control of the whole internal carotid artery circumference in the inferior and mesotympanic portion.

cochlea into the petrous apex. Hemostasis is best performed by respecting the cleavage plane on the ICA, coagulating the caroticotympanic arteries, and using a diamond burr.

In patients with poor hearing or EAC erosion, a canal wall-down procedure without FN rerouting can be performed (Fig. 3). Blind sac closure of the EAC and obliteration of the cavity

Figure 3 In patients with poor hearing or external canal erosion, a canal wall-down procedure without facial nerve rerouting is performed.

complete the surgery in this case. In the unusual event that the FN is infiltrated by the tumor, it is often possible to strip the epineurium with the tumor or, if necessary, the nerve can be cut and repaired at the end of surgery.

The sigmoid sinus is occluded by extraluminal packing with Surgicel below the superior petrosal sinus. The internal jugular vein is double-ligated and cut in the neck immediately before tumor removal to avoid unnecessary venous congestion. After opening the sigmoid sinus, the intraluminal portion of the tumor is removed, and bleeding from the inferior petrosal sinus and the condylar emissary vein is controlled with packing. Preservation of the medial wall of the jugular bulb helps to protect the lower cranial nerves in the jugular foramen.

Intradural tumor extension is addressed at the final stage after adequate hemostasis is provided by removal of the extracranial tumor component. The positions of the FN, the middle ear components, and the EAC play a subordinate role in exposure during the intradural resection. Any dural defect is covered with layers of fascia, and abdominal fat is secured with fibrin glue and covered by a temporalis muscle flap.

Ossicular chain reconstruction is performed either at the time or as a second stage procedure. The details of the IFT-A approach have been presented elsewhere.¹⁹

Follow-up

Follow-up duration ranged from 2 to 15 years (mean, 8.2 years). The postoperative investigations included serial magnetic resonance imaging (MRI), dynamic magnetic resonance contrast studies, cranial nerve (CN) functional testing, hearing tests, and angiographic studies in selected patients.²⁰

RESULTS

Complete tumor removal was achieved in all but 6 patients using the TM-IL approach (84%) and in 8 of 11 patients with IFT-A approach (73%) (Table 1). In both groups, residual tumor was found either in the region of the jugular foramen or the petrous apex medial to the petrous carotid artery.

Facial nerve function is documented in Table 2. In the TM-IL group, one patient had preoperative signs of FN involvement, but it was possible to remove the infiltrated epineurium and still preserve neural continuity. In other TM-IL patients, the FN was either skeletonized or mobilized from the second genu, resulting in postoperative House-Brackmann (HB) grade I or II in 84% of cases.²¹ In the IFT-A group, two patients had preexisting facial paralysis that required grafts.

Table 2 Pre- and Postoperative Cranial Nerve Function Results

CN VII HB Index	TM-IL (N = 38)		IFT-A (N = 11)	
	Preoperative	Postoperative	Preoperative	Postoperative
I	37 (97%)	26 (68%)	9 (82%)	5 (46%)
II	0	5 (13%)	0	0
III	0	4 (11%)	0	1 (9%)
IV	1 (3%)	2 (5%)	0	4 (36%)
V	0	0	0	0
VI	0	1 (3%)	2 (18%)	1 (9%)
CN IX Palsy	8 (21%)	11 (29%)	2 (18%)	4 (36%)
CN X Palsy	10 (26%)	14 (37%)	3 (27%)	5 (46%)
CN XI Palsy	0	4 (11%)	1 (9%)	2 (18%)
CN XII Palsy	4 (11%)	10 (26%)	2 (18%)	4 (36%)

CN, cranial nerve; HB, House-Brackmann classification; TM-IL, transmastoid-infralabyrinthine; IFT-A, infratemporal fossa type A.

House-Brackmann grade I or II FN function was obtained in 56% 1 year after surgery.

Peri- and postoperative morbidity and complications are shown in Table 2. The incidence of acquired lower CN palsies was 8% for CN IX, 11% for CN X, 11% for CN XI, and 15% for CN XII in the TM-IL group. In the IFT-A group, it was 18% for CN IX, 19% for CN X, 9% for CN XI, and 18% for CN XII. Although there were no fatalities in the whole series, the incidence of meningitis and transient cerebrospinal (CSF) fluid leak was higher in the TM-IL group (8%, 13%) compared with the IFT-A group (0%, 9%). In two TM-IL patients, revision surgery was necessary to control the leak (5.3%).

DISCUSSION

The IFT-A approach was introduced by Fisch in 1977, and his systematic description of the management of the FN in the course of jugular PGL surgery followed in 1979.^{1,22} Temporary FN displacement after removal of the posterior canal wall during jugular PGL surgery had been proposed by other surgeons previously.^{23–25} To reduce trauma to the nerve and to minimize interference with its blood supply, en bloc displacement of the stylomastoid foramen soft tissue together with the FN, and the continuous use of intraoperative FN monitoring were proposed.^{6,12} Glasscock et al described an alternative technique of jugular PGL removal with temporary FN displacement and preservation of the EAC.⁴ In an anatomical and radiological study, Maniglia et al promoted the technique of inferior mastoidectomy–hypotympanic dissection for the removal of jugular PGL, which does not invade the facial recess, attic, or mastoid antrum.⁷ The FN is partially mobilized from the inferior aspect of the vertical portion of the fallopian canal and reflected anteriorly. The integrity of the external ear canal and the tympanic membrane are preserved. Jackson addressed the problem of blind dissection of middle ear pathology from the vertical segment of the ICA,

describing the infratympanic extended facial recess approach.⁹ It affords identification of the jugular bulb; the intratympanic ICA is completely exposed to just beyond its genu, and the medial 25% of the eustachian tube is accessible. The FN is skeletonized, leaving a thin covering of bone. It is best suited to benign middle ear pathology, such as extensive glomus tympanicum tumors, and was demonstrated with a series of three patients.

Summarizing the outcomes of intratemporal FN rerouting, Parhizkar et al (2005)²⁶ produced an updated comprehensive review based on Selesnick's data (1996).²⁷ By analyzing all preceding studies from 1977 to 1996, both studies showed that patients with short anterior rerouting had the best FN function, with 47% achieving HB grade I or II function at short-term follow-up and 93% at long-term follow-up. Analysis of the series on long anterior rerouting showed that 41% of patients had grade I or II function at short-term follow-up and 73% had grade I or II at long-term follow-up.

Only a limited number of studies report long-term outcome after jugular PGL surgery, and to our knowledge no studies that compare the outcome as a function of tumor exposure gained by different strategies of FN management have been published. Although the extent of surgical exposure is dictated by such factors as perineural tumor spread within the jugular foramen, bone, and vascular areas, adequate tumor exposure is the prerequisite for safe and complete tumor removal. With the IFT-A approach, Briner et al reported that complete tumor removal after 10 to 15 years was achieved in 83% of patients.² In a series of 53 patients with Fisch class C and D tumors, Sanna et al achieved gross total tumor removal in 49 patients (90.7%), in whom recurrence was documented in a further five (10.2%) cases, making the surgical control rate 83%.³ The follow-up period was rather short, ranging from 3 to 144 months (mean, 31.3 months).

As an alternative, Pensak and Jackler described the creation of a fallopian bridge with preservation of the FN when removing tumors

that have developed within or juxtaposed to the jugular fossa.¹¹ In the study of 35 patients, of which only 13 had jugular PGLs, 71% had complete removal of tumor; subtotal removal was achieved in 20%, and partial removal was accomplished in 9% of patients. Unfortunately, they did not give precise pathological data or state the length of postoperative follow-up. The authors concluded that in selected cases, the FN should be left in place until the surgeon decides that mobilization is absolutely necessary for complete and safe resection of a given lesion. In their opinion, permanent anterior FN transposition was required for cases in which the tumor had eroded the carotid canal at the genu, whereas the presence or absence of intracranial extension had little relevance to this decision. Jackson et al reported complete surgical control in 85% of cases (155 out of 182 procedures), with an average follow-up of 54 months (range, 1 to 279 months).¹⁴ In 2004, Borba et al presented a series of 24 jugular PGL class C and D tumors, which were treated without rerouting of the FN via the infralabyrinthine approach.¹⁵ Radical tumor removal, as determined by MRI, was achieved in 23 of 24 (95.8%) patients at a mean follow-up period of 45 months. In 2004, Ramina et al presented a series of 102 jugular foramen tumors, of which 58 were jugular PGLs.²⁸ The tumor sizes were not further classified, and the follow-up interval was not stated. Radical removal was possible in 45 of the 58 patients (77.5%). Because three of these developed recurrences, the overall control by primary surgery was 72.4%. The FN was not removed from its bony canal unless infiltrated by the lesion. Tumor removal was accomplished anteriorly and posteriorly to the facial canal.

Our study compares the functional results of jugular PGL surgery achieved by two surgical techniques performed by one surgeon. The decision whether to use the TM-IL (78% of patients) or the IFT-A (22%) approach was made individually and based on the degree of destruction of the middle ear, ICA encasement, and the extent of any dural defect after resection of the intradural com-

ponent of the tumor. The TM-IL approach, as performed in our hands, is a compilation of different surgical steps developed by our own experience together with the experience of others.⁴⁻¹¹ Complete tumor removal was achieved in 84% of patients after TM-IL and in 73% after IFT-A. This can be explained by the fact that the IFT-A approach was applied to patients with more extensive disease—C3 tumors with significant carotid artery involvement (45% compared with 32% in TM-IL) and intradural tumor spread (18% compared with 8% in TM-IL). Nevertheless, the rate of 84% of tumor control achieved after an average follow-up of 8.2 years encourages us to continue to use an individually tailored FN technique via the TM-IL approach. This technique is easily converted to the IFT-A approach if additional exposure is required for safe tumor resection from the ICA or the petrous apex, or if the extent of the dural defect makes obliteration of the middle ear space mandatory. Following this principle, the postoperative FN results in our series confirm previous reports from the literature. In two patients with preoperative FN paralysis, the IFT-A approach was used and the nerve was resected and grafted at the end of surgery. In a patient with preoperative HB grade IV palsy, an epineural tumor resection was performed and the continuity of the nerve preserved. The function recovered to HB grade II.

Cerebrospinal fluid leak is a recognized complication of jugular PGL surgery. The incidence of a transient subcutaneous CSF leak after IFT-A is reported to be 14%,² and of a permanent leak requiring additional surgical revision is stated to be 3.8%.³ For surgical procedures that include the concept of middle ear preservation and a tailored FN strategy, the incidence of CSF leak varied between 3.9 and 8%.^{14,15,28} In our hands, using the TM-IL approach, a transient leak rate of 13% was experienced and only 5.3% required revision surgery. This correlated well with other series and showed that individually tailored function preservation surgery carries the same, if not smaller, risk as other procedures.

CONCLUSIONS

The TM-IL approach with individually tailored management of the FN enables safe and radical removal of the majority of jugular PGLs. It fulfills the requirements of both maintaining good post-operative FN function and obtaining an adequate tumor resection with good long-term results. Post-operative morbidity compares favorably with the IFT-A approach, particularly in respect to hearing preservation. Limitations of this approach are giant tumors with extensive anterior and pericarotid spread, which require maximal exposure for safe resection.

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