Comparison of Lower Cranial Nerve Function Between Tympanojugular Paraganglioma Class C1/C2 With and Without Intracranial Extension: A Four-Decade Experience

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Objective: To compare preoperative and postoperative lower cranial nerve (LCN) function between Class C1 and C2 tympanojugular paraganglioma (TJP) with/without intracranial intradural (Di)/extradural (De) extensions, according to the experience of a single surgeon over four decades.
Study Design: Retrospective review.
Setting: Quaternary referral center for otology and skull base surgery.
Material and Methods: A chart review was conducted of all the patients operated for C1/C2 TJPs from September 1983 to December 2018. The tumors were classified as: Limited-Group (C1/C2 without Di/De extensions) and Extended-Group (C1/C2 with Di/De extensions).
Results: Of 159 patients, 107 (67.3%) were women; the mean age at surgery was 46.5 years. The Limited-Group (56.6%) comprised C1 (41.1%) and C2 (58.9%) tumors; the Extended-Group (43.4%) comprised C1+Di/De (14.5%) and C2+Di/De (85.5%) tumors. The prevalence of preoperative LCN palsy was 11.9 times higher in Extended than Limited tumors: 61.9% versus 4.9% ($p < 0.05$). The risk for postoperative LCN palsy was 4.7 times greater in Extended than Limited tumors: 29.2% versus 12.9%, $p = 0.01$.
Conclusion: Especially in younger patients, complete removal of Limited C1/C2 tumors, before they extend intracranially, reduces the risk of dysfunctionality of LCNs and the burden of residual tumor. The incidence of new tumors increased over four decades. However, new-postoperative LCN palsy did not occur in any Limited C1/C2 tumors operated after the year 2000, and declined to less than 10% of Extended C1/C2 tumors. **Key Words:** Extradural extensions—Intradural extensions—Lower cranial nerve function—Surgical experience—Tympanojugular paraganglioma.

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Tympanojugular paragangliomas (TJPs) are highly vascular, principally indolent, benign, and slow-growing tumors. Nevertheless, their locally-aggressive behavior in a complex anatomical location, and the potential for intracranial involvement, render surgery a particularly challenging task, with possible intraoperative and postoperative complications. This necessitates adequate surgical skills, which are available only at a limited number of highly specialized centers.

Although rare, with an incidence of 8.6 per 100,000 cases, TJP account for 80% of all neoplasms at their site (1–3). TJP typically originate from the adventitia of the superolateral portion of the jugular bulb (JB) dome, and extend to gradually fill the lateral compartment of the jugular foramen. This consequently displaces medially the lower cranial nerves (LCNs), which are covered and protected by the medial wall of the JB (4,5).

In the context of the anatomical and tumoral aspects described above, the ''intrabulbar dissection technique'' has developed (6,7), which preserves the antero-medial wall of the JB, thus protecting the LCNs. This procedure is adequate, especially for early and limited C1 and C2 tumors, without medial wall invasion, for preservation of intact LCN function (4,6,8–10).

Despite the above, once these tumors transgress the medial-wall area dura, though still classified as C1/C2, the risk for abnormal LCN function increases. This produces a burden of dysphonia, dysphagia, and aspiration (11–16), which for younger patients is mostly transient, curable, and acceptable (17–19); while in older persons, such abrupt LCN dysfunction can be fatal (18–22).

To the best of our knowledge, no reports on LCN function comparing TJP Classes C1 and C2, with or

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without dural extension, have been published in the English literature. The aim of the present study was to compare preoperative and postoperative LCN functional status, between early limited C1/C2 Classes and their counterparts with intracranial dural extensions. The outcomes of four decades of experience were compared with the existing literature.

MATERIALS AND METHODS

After institutional review board approval, the records of all patients treated at our institution with Fisch Class C and D TJP (20) from September 1983 to December 2018 were analyzed. Only those with tumors diagnosed as Class C1 and C2 with/without intracranial extensions according to preoperative imaging were included in this study. Patients with incomplete records or lost to follow up for a minimum period of 1 year were excluded from the analysis. The cohort was stratified by tumor classification: ''Limited Group'' (C1/C2 without Di/De extensions) and ''Extended Group'' (C1/C2 with Di/De extensions).

Patients with simultaneous carotid/vagal paragangliomas, who had preoperative LCN palsy, were excluded from the LCN palsy analysis, due to the inability to discern whether their palsies were due to TJP or to other harbored tumors.

We stratified the data by the abovementioned groups, to examine tumoral distribution and incidence. To evaluate the impact of the personal surgical experience throughout four decades, we established three time-frame periods: early (1983–1995), intermediate (1996–2007), and late (2008–2018). All the patients underwent complete preoperative otoneurologic evaluation. Glossopharyngeal nerve function was assessed by checking for the presence of gag reflex. Vagal nerve function was assessed by checking for soft palate motility and vocal fold movement. A fiberoptic laryngoscopy was performed both preoperatively and postoperatively. This examination may confirm nerve injury in the presence of vocal fold weakness, which may influence dysphagia and dysphonia.

The patients were asked to raise both their arms above their head to assess accessory nerve function, and were checked for tongue deviation to assess hypoglossal nerve function. Facial nerve function was graded preoperatively and postoperatively by the House–Brackmann grading system (23), and hearing was assessed by pure-tone audiometry.

All the patients underwent thorough preoperative imaging evaluation, which consisted of high-resolution computed tomography (CT) with reconstructions in the axial and coronal planes to identify the exact extent of air-cell involvement. TI-, T2-, and gadolinium-enhanced T1-weighted magnetic-resonance-imaging (MRI) was required to distinguish TJPs from other jugular foramen lesions, and to assess the intracranial and intradural extensions.

Angio-MRI, angio-CT, and four-vessel angiography with manual cross-compression testing were also performed to elucidate tumor vascularization and to plan preoperative embolization, as well as to assess internal carotid artery involvement and contralateral venous drainage. Operations were done within 72 hours after embolization to preclude development of collateral feeding vessels and the development of post-embolization inflammatory effects.

An infratemporal-fossa approach type A was employed in all the patients. The surgical steps have been described elsewhere (24–26). Transcondylar-transtubercular extension for obtaining posteroinferior and medial access to the JB was employed as was appropriate in patients with C2 tumors. A modification to the Fallopian bridge-technique was employed in two patients who presented with Class C1 tumors and predominantly posterior disease. Facial nerve monitor was applied in every patient; however, the LCNs were not monitored intraoperatively.

Postoperative LCN function was assessed at 3 months and at the last follow-up (1-year minimum) by means of flexible video-endoscopy with functional evaluation of swallowing and sensory testing.

Postoperative follow-up MRI was performed at the third and twelfth months and subsequently once every year. The tumoral-residue (in mm) was measured in two perpendicular directions and a growth rate of greater than 3 mm/yr was considered a fast-growing residue, while less than 3 mm/yr was considered a slow growing residue (27).

Statistical Analysis

The data were analyzed using SPSS 24 (IBM, New York, NY) statistical package. The χ^2 test was used to measure the significance for the nonparametric data. A p-value <0.05 was considered statistically significant (95% confidence interval).

RESULTS

A total of 267 patients with TJP were treated during the study period. For 52 (19.5%), a wait-and-scan strategy was adopted. Of the 215 patients who underwent surgery, the 159 (59.5%) with TJPs assigned as Classes C1 and C2, with/without intracranial extensions, comprised the study cohort. The mean follow-up was 78.1 months (median, 59.2; range, 12–341).

Demographics and Symptomatology

The mean age of the study population at the time of surgery was 46.5 years (median 46; range, 12–71); 107 (67.3%) were women. Reddish retro-tympanic pulsative mass was the most common presenting sign, appearing in 123 (77.4%) patients. Pulsatile tinnitus presented in 111 (69.8%) patients and 122 (76.7%) patients cited hearing loss. Among those with LCN deficits, dysphonia was the most commonly reported symptom, presenting in 43 (27%) patients (Table 1). Neither malignancy nor catecholamine-secretion was observed in any of the patients. The mean duration of symptoms before seeking medical attention was 2.9 years.

Seventeen patients synchronously harbored benign, non-secreting tumors: carotid body, vagal, and simultaneous carotid/vagal paragangliomas were revealed in nine, six, and two patients, respectively. All the tumors were concomitantly surgically treated.

Distribution of Tumors (n = 159)

The Limited Group comprised 90 (56.6%) patients: 37 (41.1%) C1 and 53 (58.9%) C2. Of the 69 (43.4%) patients in the Extended Group, 10 (14.5%) had Class C1 and 59 (85.5%) C2 tumors, with D extensions. Extradural and intradural involvement was higher in Class C2 tumors: 23 (20.5%) and 36 (32.1%), respectively, than in Class C1 tumors: five (10.6%) and five (10.6%), respectively. However, only the difference in

TABLE 1. *Demographics and symptomatology of the total cohort (n = 159)*

Age, years, mean, (median, range)	46.5, (12–71)
	n (%)
Gender	
Male	52 (32.7)
Female	107 (67.3)
Laterality	
Right	73 (45.9)
Left	86 (54.1)
Multicentric tumors	17 (10.7)
Signs and symptoms	
Normal otoscopy	25 (15.7)
Red retrotympanic pulsatile mass	123 (77.4)
EAC inferior wall mass	18 (11.3)
Red pulsative tumor in the CWD cavity	1 (0.6)
Otorrhea	6 (3.7)
Aural fullness	3 (1.8)
Normal hearing	26 (16.4)
Hearing loss	122 (76.7)
Anacusis	14 (8.8)
Pulsatile tinnitus	111 (69.8)
Non-pulsatile tinnitus	29 (18.2)
Dizziness/Vertigo	27 (16.9)
Dysphagia	35 (22.0)
Dysphonia	43 (27.0)
Facial weakness	25 (15.7)
Facial hypoesthesia	1 (0.6)
Shoulder pain	16 (10.1)
Tongue deviation	12 (7.5)
Upper cervical mass	1 (0.6)
Follow up, mean months, (median, range)	78.1, (12–341)

CWD, Canal Wall Down; EAC, External Ear Canal.

occurrence of intradural extensions between the tumoral classes met statistical significance, OR = 3.97, $p = 0.01$ (Table 2). The distributions of tumoral classes across the three time-intervals were not significantly different (Table 2).

Distribution of Tumors According to the Presence of LCN Palsies (n = 159)

Limited Group (C1 and C2 Classes) (n = 90)

Of 37 patients with Class C1 tumors, three patients who had preoperative LCN palsies and simultaneous carotid/vagal paraganglioma were excluded from this analysis. One patient had preoperative IX/X palsy. Postoperatively, new palsies were observed in four of 34 (11.7%) patients; two revealed both IX/X and XI palsy, one had IX palsy, and the remaining patient had X palsy. No change was observed postoperatively in the patient with preoperative IX/X palsy. All these patients with postoperative palsy were operated during 1983 to 1991.

Of 53 patients with Class C2 tumors, nine had preoperative LCN palsies. Two of these revealed X palsy, and one had IX palsy. Another five patients with simultaneous carotid/vagal paragangliomas and one patient who was operated elsewhere and presented with postoperative palsies and a remnant tumor were excluded from this analysis. Postoperatively, in six of 47 (12.7%) patients, new palsies were observed: IX palsy in two patients, IX/X palsy in another two, and IX/X/XI palsy in two patients. No change was observed postoperatively in the three patients with preoperative palsies. All these patients with postoperative palsy were operated during 1983 to 1999.

Occurrences of new postoperative palsy were not more common in those with C1 than C2 (OR = 1.085, $\chi^2 = 0.03$). However, analysis of the Limited Group as a whole shows that 10 (12.9%) of the 77 with intact preoperative LCN function developed new transient postoperative palsy. This observation translates to a 3.3 times higher relative risk for a new postoperative palsy in this group, though this association was not statistically significant ($\chi^2 = 2.38$).

Tumoral incidence increased over the study period: the mean numbers of annual incidences were: 2.4, 6.0, and 6.4 for the early, intermediate, and late periods, respectively. However, the tendency for the development of

TABLE 2. *Distribution of Limited and Extended groups across the time intervals (n = 159)*

	Early 1983–1995 (n = 29)	Intermediate 1996–2007 (n = 66)	Late 2008–2018 (n = 64)	Total (n = 159)
Class		n (%)		
Total C1 = 47				
C_1	7 (24.1)	12 (18.2)	18 (28.1)	37 (23.3)
C_1De_1*	2 (6.9)	1 (1.5)	2 (3.1)	5 (3.1)
$C_1Di_1/Di_{2\#}$	0 (0)	3 (4.5)	2 (3.1)	5 (3.1)
Total C2 = 112				
C_2	9 (31.0)	22 (33.3)	22 (34.4)	53 (33.3)
C_2De_1/De_2	3 (10.3)	14 (21.2)	6 (9.4)	23 (14.5)
$C_2Di_1/Di_2/Vi$**	8 (27.6)	14 (21.2)	14 (21.8)	36 (22.6)

Abbreviations and explanations: *: ''e'' stands for extradural extensions, with 1 or 2 for up to 2 cm or more than 2 cm, respectively. #: ''i'' stands for intradural extensions, with 1 or 2 for up to 2 cm or more than 2 cm, respectively. **: Vi stands for intradural vertebral artery involvement, occurring in four of C2Di2 cases.

TABLE 3. *LCN palsy between Limited and Extended groups across the time intervals (n = 159)*

LCN Palsy		Early 1983–1995 (n = 29)	Intermediate 1996–2007 (n = 66)	Late 2008–2018 (n = 64)
C_1+C_2		(n = 14)*	(n = 31)#	(n = 36)§
	Preop n (%)	2 (14.3)	0 (0)	2 (5.6)
	Total postop n (%)	9 (64.3)	3 (9.7)	2 (5.6)
	New postop n (%)	7 (50)	3 (9.7)	0 (0)
(C_1/C_2) + D		(n = 13)	(n = 26)¤	(n = 24)
	Preop n (%)	10 (76.9)	15 (57.7)	14 (58.3)
	Total postop n (%)	12 (92.3)	18 (69.2)	16 (66.6)
	New postop n (%)	2 (15.4)	3 (11.5)	2 (8.3)

Abbreviations and explanations: *: One patient was previously operated elsewhere and had LCN palsy postop., the other had simultaneous Carotid body tumor (CBT). Both were excluded.

#: Three cases had vagal paraganglioma (PG) or CBT preop. and were excluded. §: Four out of six had simultaneous vagal PG/CBT preoperatively and LCN palsies and were excluded.

¤: Out of 32, six had simultaneous CBT/vagal PG and LCN palsies preop., and were excluded.

new postoperative palsies decreased: from seven to three to zero palsies for the respective periods (Tables 2 and 3).

Extended Group (Class C1 and C2 with De/Di/Vi Extensions) (Total n = 69)

After excluding six patients with synchronous carotid/vagal paragangliomas, preoperative palsies were observed in 39 of 63 (62%) patients. Eighteen patients had IX/XI palsies, 10 had IX/X/XI/XII palsies, five had IX/X/XI palsies, four had IX palsy, and one each had X/XI/XII and IX/X/XII palsy.

Of 24 patients with normal preoperative LCN function, 7 (29%) developed new postoperative palsy. The relative risk for developing new postoperative palsy was 4.7 times higher in this group; the association was statistically significant ($p = 0.01$, $\chi^2 = 7.65$) (Table 3). The incidence for developing postoperative palsies was 3.6 times greater in the Extended (7/24, 29%) than the Limited Group (10/77, 13%), $p < 0.05$, $\chi^2 = 4.81$.

Expectedly, the incidence of preoperative LCN palsy was greater, by 11.9 times in the Extended (39/63, 62%) than the Limited Group (4/81, 5%), $p < 0.05$, $\chi^2 = 6.08$ (Table 3, Fig. 1). Though the incidence of newly discovered TJP tumors has remained stable over the last four decades, a decreasing trend for new postoperative palsies has been observed (Fig. 1).

Details of LCN Impairment in the Extended Group (n = 63)

Preoperatively, as well as postoperatively, the most commonly affected nerves were the glossopharyngeal: 38 (60%)/37 (59%) and the vagal nerve: 46 (70%)/43 (65%).

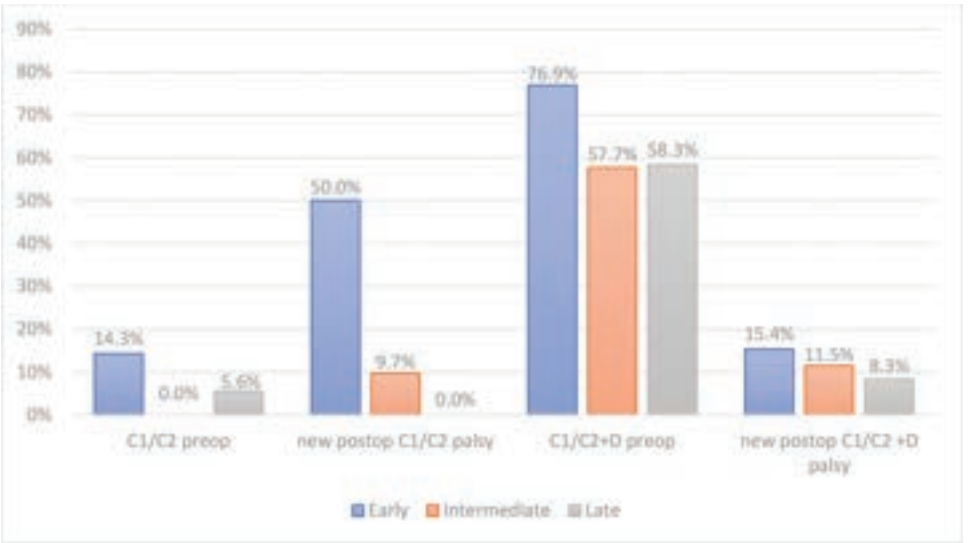

FIG. 1. In the Limited Group (C1/C2 Class tympanojugular paraganglioma without Di/De extensions), a difference is noted between the early and intermediate/late periods regarding occurrence of new postoperative palsy. The last postoperative palsy in this group occurred before the year 2000. In the Extended Group (C1/C2 Class tympanojugular paraganglioma without Di/De extensions), a decreasing trend for the new postoperative palsies is observed across the three time-intervals.

New LCN palsies were observed in a lower proportion of extradural than intradural extensions: 11 (41%) versus 20 (56%), $p < 0.05$, $\chi^2 = 5.26$.

Complications and Recurrences

The most common postoperative complication was cerebral spinal fluid leakage, which occurred in six (3.7%) patients who were successfully treated by compressive bandage. One patient with a Class C2Di2 tumor who developed a new IX–XII palsy underwent tracheotomy due to respiratory distress.

Permanent dysphonia was observed in five (3.1%) patients. In two patients (both with C2-De1 tumors), dysphonia was successfully treated by Teflon-injection medialization; satisfactory contralateral compensation was observed at 1-year postoperative. Another two patients (with C2-Di1 and C2-De1 tumors) underwent thyroplasty type 1 procedures at 1-year postoperative, with favorable outcomes. The remaining patient with a C2De2 tumor had a successful VOX implantation in the fifth postoperative year.

Transitory dysphagia occurred in two patients (1.3%) with C2 tumors, and resolved completely in 3 to 6 months. In one patient with a C2-Di1 tumor, dysphagia persisted at 1-year follow-up and was subsequently managed by a gastrostomy. All the patients with new XI palsy were satisfactorily managed by physical therapy.

Twenty-one patients (13.2%) underwent subtotal tumoral resection due to intracranial involvement. This included patients with posterior fossa dura invasion who had cuff of tumoral tissue remaining around functioning LCNs, and vertebral artery invasion.

Tumoral recurrence was observed in four (2.5%) patients (two with C1Di2 tumors and two with C2-Di2 tumors) after a mean 4.5 postoperative years. Since this was a stable finding, we opted for a wait-and-scan strategy for these patients.

DISCUSSION

Given its slow growth and the nonspecific nature of the complaints, presentation of TJP is, on average, 2 to 3 years in delay of symptom onset (5,28). Meanwhile, the tumor may reach a considerable size, thus resulting in important management issues regarding preservation of the LCNs during surgery (18).

Eighty-seven percent of the patients of our cohort with early, limited C1 and C2 tumors, with intact LCNs, maintained normal LCN function after total gross resection. The relatively young age of the patients, mean 46.5 years, poses an important consideration, as in younger patients, postoperative dysfunctional issues tend to disappear within a few weeks to months (17–19,29,30). The explanation for the preservation of LCN function lies in both the convenient anatomical features and the tumoral aspects. Since limited C1 and C2 lesions occupy only the lateral portion of the JB, they facilitate the use of the ''intrabulbar dissection'' technique (6,7,31). Accordingly, the anteromedial wall is maintained (24), thereby not jeopardizing the functionality of underlying LCNs (32) (Figs. 2 and 3). Preservation of LCN function is also facilitated by early identification in the upper neck (5,33,34) (Fig. 3).

In contrast to the above, for C1 and C2 tumors with intracranial dural transgression, our findings showed that

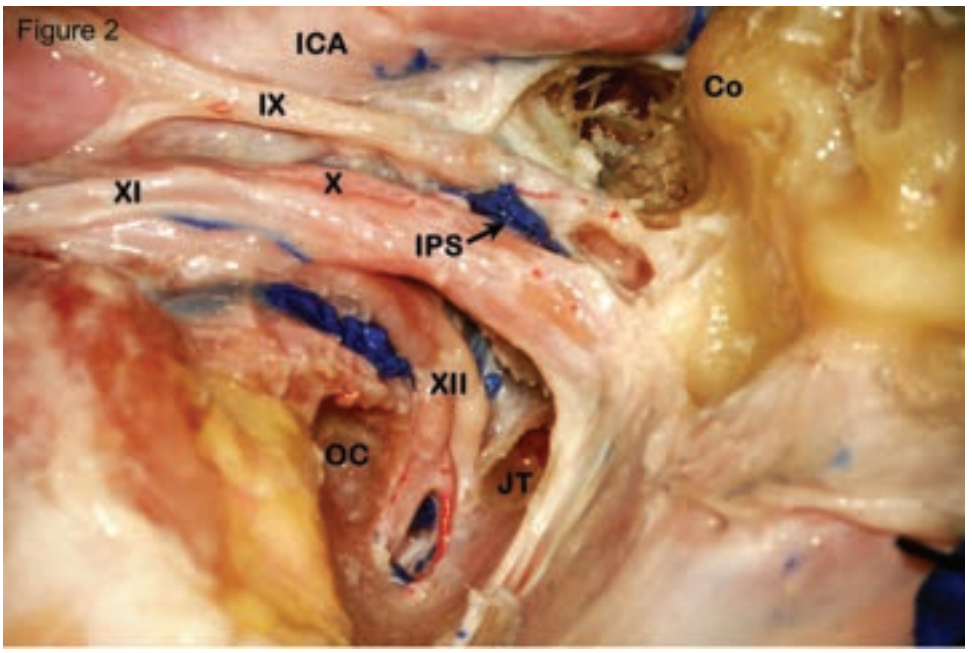

FIG. 2. *Left*-side specimen. A panoramic view of all four lower cranial nerves (LCNs) after removal of the occipital condyle (OC) and jugular tubercle (JT). (XII) Hypoglossal nerve; (IX) glossopharyngeal nerve; (X) vagus nerve; (XI) accessory nerve. The inferior petrosal sinus (IPS) can be appreciated between the IX and X nerves. (Taken with permission of Stuttgart, NewYork: Thieme, [2018]. The Temporal Bone: Anatomical Dissection and Surgical Approaches).

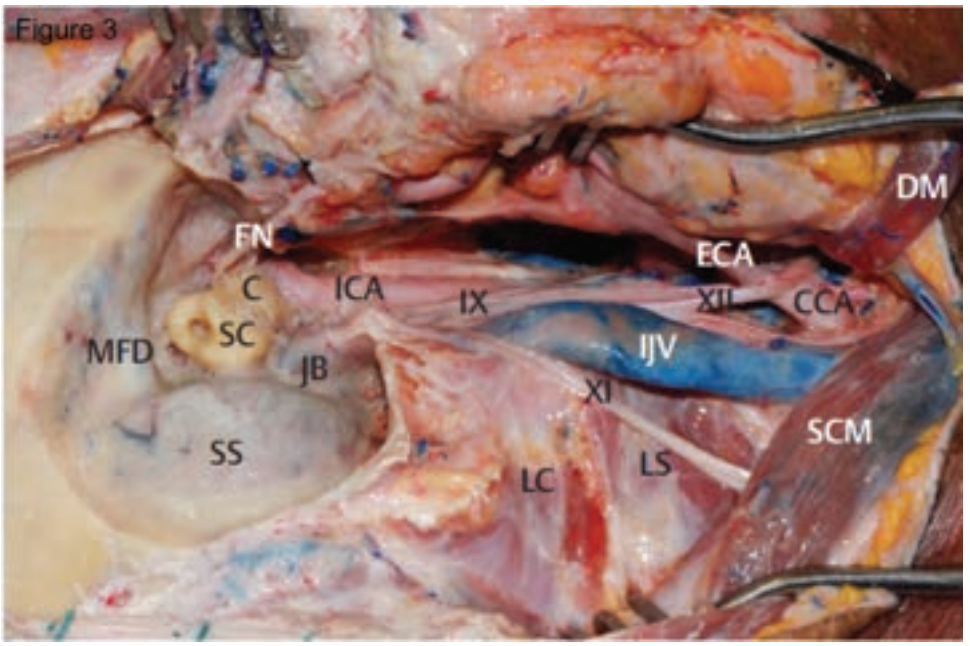

FIG. 3. *Right*-side specimen. After removal of the styloid, the internal carotid artery (ICA) can be controlled from the neck to the skull base. C indicates cochlea; CCA, common carotid artery; DM, posterior belly of the digastric muscle; ECA, external carotid artery; FN, facial nerve (rerouted); IJV, internal jugular vein; IX, glossopharyngeal nerve; JB, jugular bulb; LC, longissimus capitis muscle; LS, levator scapulae muscle; MFD, middle fossa dura; SC, semicircular canal; SCM, sternocleidomastoid muscle; SS, sigmoid sinus; XI, accessory nerve; XII, hypoglossal nerve. (Taken with a permission of Stuttgart, NewYork: Thieme, [2018]. The Temporal Bone: Anatomical Dissection and Surgical Approaches).

postoperative LCN palsy occurred in every third patient (29.2%). Here, invasion of the medial wall of the JB is crucial: when the tumor reaches this area, removal of the entire bulb and sacrifice of LCNs becomes unavoidable if the objective is to achieve oncological radicality.

Another important factor in the preservation of LCNs is the degree of tumoral occupation of the inferior petrosal sinus (18). This channel may have multiple branches, and these usually pass between the IX nerve and X/XI nerves (32,35) (Fig. 2). Consequently, care must be taken during sinus overpacking or excessive coagulation (9,24,32,36) to avoid pressure or thermal damage to the nerve fibers.

Jansen et al. (37) reported that the timing of treatment initiation represented the greatest challenge and was actually an independent predictor of treatment outcome, as once tumor growth occurred, the chance for symptom recovery diminished. Furthermore, when dealing with larger tumors with intact LCN function, Harati et al. (29) advocated a wait-and-scan approach or primary radiation, since the recovery of LCN deficits would not be expected.

In our center, we considered preoperative preservation of LCN function in patients aged over 65 years to be a contraindication for surgery. This is because compensation following acute compound palsy is particularly difficult in older persons (19), as has been described by others (18,20,21,38). A wait-and-scan strategy seemed more fitting in such context, and patients were referred to radiotherapy if there was evidence of tumor growth. Primary radiation can be considered in patients with comorbidities or poor general health (27), as has been described by others (39–44).

As previously reported (27), primary radiation, as well as a wait-and-scan policy, are not recommended in younger patients, because managing the original or recurrent tumor becomes increasingly difficult over time. Thus, for younger patients, an attempt to completely remove the tumor remains the mainstay strategy (17,33,36,39,45–50).

We report a risk for preoperative palsy that was 11.9 times higher among C1/C2 tumors with than without Di/De extension: 61.9 versus 4.9%. Furthermore, the risk for newly obtained dysfunction was 3.6 times higher in the former than the latter: 29.2% versus 12.9%. Indeed, the relative risk for developing a new postoperative palsy was 4.7 times higher among those with than without Di/De extension.

Additionally, in both C1 and C2 tumors without Di/De extension, occurrences of a new palsy were transient and similar, 11.7 and 12.7%, respectively. Thus, the extension of C1 tumors by a few more millimeters towards the genu of the internal carotid artery, which is then classified as a C2 tumor, made no practical difference in postoperative LCN status in the absence of Di/De extension. Nevertheless, the enhanced propensity for intracranial extension should be kept in mind, especially intradural extension, as tumoral class rises (10). In our cohort, intradural extension was three times higher in patients with C2 (32.1%) versus C1 (10.6%) Class tumors. Our observations corroborate reports (5,29) that

demonstrated a relation between the risk for postoperative palsy and severe intradural infiltration.

Interestingly, Sen et al. (31) found that normal preoperative LCN status does not eliminate tumor infiltration. Indeed, preoperative compensated ''silent'' LCN deficits were shown to occur in up to 10% (51,52). In this context, sacrificing such nerves severely worsens LCN function, probably due to the dissection of residual functioning fibers that maintain a degree of tonus (18,24). Accordingly, we left a small cuff of tumor around nerves that were found to infiltrate but still function, as did others (5,29), or that risked LCN dysfunction by gross total resection. Clinical decisions were based on the natural tumor history, age, general health status, and mutual preoperative discussion, as described by others (29).

Postoperative LCN dysfunctionality seems inevitable following the surgical removal of tumors with an intradural component. In five of 24 patients in one series (53), the tumor involved the neural compartment, and was thus treated by conservative resection and postoperative radiation; normal LCN function was achieved. Karaman et al. (40) reported that preoperative LCN palsy was encountered in every tumor with intracranial extension, and in only two of nine limited tumors. In yet another study (41), penetration of the tumor into the neural compartment within the jugular foramen resulted in full postoperative LCN palsy in all 21 patients, even in those without preoperative deficits. Considering the current findings, together with the abovementioned reports from the literature (5,29,40,41,53), preservation of LCN function seems impossible once a tumor becomes intradural. Thus, decisive surgical treatment appears as an effective strategy, while the tumor is still considered small and limited. The aim is to achieve complete gross dissection with minimal morbidity before any intracranial invasion occurs, when feasible. This has been shown by several reports (17,33,36,39,45–50).

Among the tumors without Di/De extensions, the 12.9% rate of postoperative dysfunctional LCNs translated to a 3.3 times higher relative risk for obtaining a new transient postoperative palsy; however, this association was not statistically significant. Furthermore, the occurrence decreased during the study period, and was nil after the year 2000. This was despite the increased tumoral incidence observed over the decades of the study (Fig. 1, Tables 2 and 3). Similarly, while the number of tumors with Di/De extensions remained relatively stable during the study period, the occurrence of a new postoperative palsy decreased (Fig. 1, Tables 2 and 3). This could be explained by improvements in intraoperative reasoning, and the development of surgical skills over decades of experience.

The main surgical tactic significantly depends on the surgeon's experience (5) and consists of completely encompassing the tumoral borders, proper early exposure, and consequent optimal control of the neurovascular structures before commencing with tumoral removal. In our cohort of C1/C2 Class tumors, for all those without Di/De extensions and for the majority of those with extensions and with superolateral JB involvement, the infratemporal fossa approach proved sufficient. This was done with the addition of transcondylar-transtubercular extension for obtaining posteroinferior and medial access to the JB. While intraoperative electrophysiological monitoring is an important surgical supplement for diminishing iatrogenic neural damage, it was not considered in the current analysis, due to its inaccessibility in the initial period of the study.

Our protocol provided satisfying outcomes: the gross total resection rate was 138 (86.8%), the recurrence rate was 4 (2.5%), and the rate of LCN dysfunction was 17 (10.7%), of which only seven (4.4%) were permanent. The glossopharyngeal nerve was found to be the most vulnerable, presumably due to its lateral positioning in the jugular foramen (4,31,32,54) (Fig. 2).

Review of the literature reveals shortcomings in the terminology of the Fisch classification for TJP (5,22,29,36,41,42,43,55,56), in that Classes C and D are referred to only broadly. This is because Class C tumors can be with or without dural extension; and vice versa, Class D tumors generally possess C components. Thus, comparison of the data is hardly possible.

Our gross-total resection rate of 138 (86.8%) is within the range of 62.5 to 95.8% reported by studies that include heterogenous tumor stages (5,20,29,36,41,42,44). We did not find data in the literature regarding the recurrence rate of Class C1/C2 tumors, for comparison with our data. Postoperative LCN palsy has been reported in a range of 4.2 to 23.5% (5,29,36,40,42); however, again, due to the variability of reported tumor classes, the comparison was not feasible.

Long-term data of TJP have accumulated, due to the growth rate of 0.8 mm/yr, translating to a tumor doubling-time of about 10 years (median 4.2 yr) (57), together with growing experience with the radiotherapy modality. In Winford et al.'s review (55), four of 14 series of patients treated with stereotactic radiosurgery for TJP showed tumor control rates of 93 to 100% during follow up periods of 50.4 to 86.4 months. Moreover, Ibrahim et al. (58) reported similar tumor growth control rates for median radiological and clinical follow-up periods of 51.5 and 38.5 months, respectively. Notably, the majority of radiotherapy series analyzed tympanojugular/tympanomastoid paraganglioma classes together, thus including a range of tumor volumes (4.14–14.20 cm^3) (55). In contrast, the current study analyzed and compared distinctive TJP Classes (C1/C2 versus C1/C2 +Di/De). This difference between studies poses difficulty in comparing the two modalities. Furthermore, Class B tympanomastoid paragangliomas (glomus tympanicum) comprised 13% of all the radiated tumors reported by Winford et al. (55). In a single-center series, 24.9% of all the irradiated tumors belonged to Class A and B paraganglioma (58). In a multicenter study (59), 8.9% of the tumors were of Class A and B; and for 4.5% of the patients, data of tumor class were missing. Tympanomastoid paraganglioma (glomus tympanicum) tumors can be completely removed by standard approaches such as radical mastoidectomy with or

without a blind-sac closure (60). Accordingly, there are practically no sequelae and patients receive a complete and permanent cure with intact inner ear function (20,60).

Pre- and posttreatment results, specifically of post-radiotherapy LCN status, are not conclusive across radiotherapy studies; a variety of tumors were included in the studies, and the specific cranial nerves involved were generally not specified (55,58,59,61–64). In three studies, however, LCN status was clearly described. Gerosa et al. (63) and Eustacchio et al. (64) reported post-radiation LCN functional improvement in 5 and 21%, respectively; while Dobberpuhl et al. (65) reported stability after radiation in seven of 12 patients with pre-treatment lower cranial dysfunction. Further studies, comprising more uniform populations of tympanojugular tumor classes, with larger cohort numbers, longer follow up, and more specific descriptions of the cranial nerve status are necessary for better comparison of data between radiotherapy and surgery.

Five of our patients experienced dysphonia after surgery, which persisted at 1-year follow-up. Three patients treated for Class C2 TJP with persistent dysphagia 1 year after surgery were managed with a gastrostomy for temporary feeding and rehabilitation of swallowing function. Many procedures have been advocated for managing LCN deficits, including tracheostomy, gastrostomy, vocal cord manipulation, and cricopharyngeal myotomy. Short-term nasogastric feeding is occasionally required, and a temporary percutaneous gastrostomy tube is inserted if swallowing problems persist for more than a few weeks. Vocal fold medicalization should be used in patients with persistent vocal deficits, using materials such as Silastic, Goretex, Teflon, and hydroxyapatite. Arytenoid adduction can be combined with a type I thyroplasty in patients with inadequate posterior closure of the glottic plane (24).

CONCLUSION

The findings of this study support the performance of complete tumor removal in small, limited C1/C2 tumors. This is especially important in younger patients, when feasible, before the tumor extends intracranially, which vastly reduces the possibility for proper functionality of LCNs, and also carries a burden of residual tumor.

We report decreased occurrence, to nil, of new post-operative LCN palsy among limited C1/C2 tumors. Moreover, we report a decline in postoperative LCN palsy to less than 10% among patients with extended intracranial C1/C2. These trends occurred in parallel to stable or somewhat increasing incidence of TJP. Good intraoperative judgment and improved surgical skills may have contributed to the positive results.

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