Original Paper

Audiology & Neurotology

Audiol Neurotol 2012;17:243–255
DOI: 10.1159/000338418

Received: November 7, 2011
Accepted after revision: March 7, 2012
Published online: May 15, 2012

Surgical Management of Tympanojugular Paragangliomas with Intradural Extension, with a Proposed Revision of the Fisch Classification

Shailendra Sivalingam[a] Masaya Konishi[c] Seung-Ho Shin[d]
Raja Ahmed R. Lope Ahmed[b] Paolo Piazza[e] Mario Sanna[f, g]

[a]Department of Otorhinolaryngology – Head and Neck Surgery, University of Malaya, Kuala Lumpur, and [b]Department of Otorhinolaryngology – Head and Neck Surgery, International Islamic University, Kuantan, Malaysia; [c]Department of Otorhinolaryngology – Head and Neck Surgery, Kansai Medical University, Moriguchi, Japan; [d]Department of Otorhinolaryngology – Head and Neck Surgery, CHA University, Seoul, Korea; [e]Department of Neuroradiology, University of Parma, Parma, [f]Gruppo Otologico, Piacenza, and [g]ENT Department, University of Chieti, Chieti, Italy

Key Words
Tympanojugular paragangliomas · Intradural extension · Staged surgery · Fisch classification

Abstract
Background: Tympanojugular paragangliomas (TJPs) with intradural extension can be successfully treated by a single or staged procedure with low surgical morbidity. ***Objectives:*** To present the clinical findings and treatment methods used for surgically treating TJP with intradural extension, as well as to discuss the complications of treatment and the relative merits of single versus staged surgery by using a comprehensive literature review comparing objective outcome measures. ***Study Design:*** A retrospective case review of 45 cases of TJP with intradural extension. ***Setting:*** A quaternary skull base and neurotologic center. ***Materials and Methods:*** The charts of 45 patients with Fisch classification class C or D TJP with intradural extension, who were operated on from April 1988 to April 2010, were analyzed. Clinical findings and preoperative lower cranial nerve (LCN) palsy as well as postoperative totality of resection, postoperative LCN palsy and complications were studied. The types, indications, and distribution of staged procedures were also analyzed. ***Results:*** Out of 45 cases, 22 were C3di2. The IX and X cranial nerves were the commonest nerves affected preoperatively. Preoperative internal carotid artery management was performed in 16 cases. Twenty-nine cases had a single procedure and 16 had a staged procedure. The main indication for staged procedures was intradural extension of 2 cm or more. The infratemporal fossa approach (ITFA) type A was the main procedure in all cases. Overall, total resection was achieved in 68.8% of cases with postoperative cerebrospinal fluid leak in 4.4% cases. Postoperative House-Beckmann grade I–III facial nerve status was maintained in 80% of cases, and overall LCN preservation rate was 56.9%. There were no cases requiring tracheostomy, and 3 cases required delayed phonosurgical procedures to improve their voice. ***Conclusions:*** TJP with intradural extension can be successfully managed with the judicious use of staged procedures to reduce the incidence of postoperative cerebrospinal fluid leak. The ITFA did not cause an excessively high rate of facial nerve palsy, and the overall total resection and LCN preservation rate compares very favorably with previously published data.

This study was conducted at the Gruppo Otologico, Piacenza, Italy.

KARGER

Fax +41 61 306 12 34
E-Mail karger@karger.ch
www.karger.com

1420–3030/12/0174–0243$38.00/0

Accessible online at:
www.karger.com/aud

Shailendra Sivalingam
Department of Otorhinolaryngology – Head and Neck Surgery
University of Malaya
Kuala Lumpur 50603 (Malaysia)
Tel. +60 122 586 047, E-Mail shailensivalingam@yahoo.com

Introduction

Tympanojugular paragangliomas (TJPs) are the second commonest tumor of the temporal bone and the commonest tumor affecting the jugular foramen [Sanna et al., 2004a]. They arise either in the adventitia of the jugular bulb or along the course of Jacobsen's nerve or Arnold's nerve. The growth of TJPs is slow and insidious, often invading and eroding the bony skull base, infiltrating the regional cranial nerves (CNs) and constricting the major blood vessels to the brain either by encasement or direct infiltration, and sometimes also transgressing the dural barrier [Sanna et al., 2004a].

The Fisch classification has been a widely accepted classification for TJP and is useful for categorizing the extent of tumors [Fisch, 1988]. The incidence of intradural extension becomes more frequent as the Fisch C class stage increases [Patel et al., 1994; Sanna et al., 2004a]. Surgery for these tumors can be very challenging, requiring the ability to assess varied interrelated factors and integrate them into the surgical plan. The larger the intradural extension is, the more frequently they involve the lower cranial nerves (LCNs) and compress the brain stem. The degree of dural involvement can also be difficult to assess both preoperatively and intraoperatively, and the likelihood of dural infiltration increases in parallel with the intracranial volume. Intracranial invasion, should it be extradural (class De) or intradural (class Di), mandates for similar surgical strategy [Sanna, 2008], with the Di component being the most relevant for planning purposes.

It has been advocated that with a multidisciplinary approach and meticulous closure, resection should be performed in a single stage regardless of the size of the intradural component [Sekhar and Oliveira, 1999; Al-Mefty and Teixeira, 2002; Jackson et al., 2004; Oghalai et al., 2004; Ramina et al., 2005]. However, sacrifice of LCNs at surgery can cause severe postoperative aspiration, and the resulting increase in thoracic pressure during cough efforts elevates intracranial pressure possibly giving rise to cerebrospinal fluid (CSF) leaks [Fisch, 1982]. We prefer a staged operation for tumors with more than 2 cm intradural extension (Fisch classification Di2) to prevent postoperative CSF leaks, and one-stage removal for tumors with 2 cm or smaller intradural extension (Fisch classification Di1) [Fisch et al., 1984; Sanna, 2008].

In our experience, the cleavage between tumor and brain stem can be easily established due to the devascularization of the tumor after the first-stage surgery and subsequent shrinkage of the intradural mass. In the current study, we report a retrospective analysis of surgical management of 45 patients with a diagnosis of a TJP with intradural extension.

Materials and Methods

The charts of 156 patients with Fisch classification class C or D TJP [Fisch, 1988] surgically treated during the period from April 1988 to April 2010 were studied and 55 patients with intradural extension were selected for this study. Four patients were unsuitable for surgery and 51 patients had surgery, performed by the senior author (M.S.). Postoperatively, 5 cases were lost to follow-up and 1 case had less than 1-year follow-up, and these cases were excluded from the study. The perioperative data and follow-up records of the remaining 45 cases were reviewed. Each patient had undergone complete radiologic examination, which includes high-resolution computed tomography with bone windows, magnetic resonance imaging (MRI) with gadolinium enhancement, and four-vessel angiography with manual cross-compression testing. All patients had at least one hearing assessment in the form of pure-tone audiometry. The facial nerve (FN) function was recorded using the House-Beckmann (HB) grading system [House and Brackmann, 1985] at each visit, and this assessment was always done by a member of the operating team.

The intraoperative records included detailed notes of the procedure, exact location and extent of the tumor, along with a record of the management of the FN and internal carotid artery (ICA). The FN monitor has been routinely used in all skull base cases at Gruppo Otologico since 1994. We do not routinely use LCN monitoring at our center because we feel its usefulness is yet to be fully established.

All tumors underwent embolization using polyvinyl alcohol 1–2 days before the procedure. Stenting of the ICA was recommended if the preoperative arteriography showed clear-cut involvement of the arterial wall. However, in previously operated or irradiated cases with sufficient collateral circulation, permanent balloon occlusion (PBO) was applied. Follow-up was defined as the period of time from last surgery to the most recent office visit. First follow-up with MRI was at 6 postoperative months to obtain a scan free of any inflammatory changes to be used as a reference. Subsequently, yearly imaging examination was performed in the absence of recurrence or residual tumors. In the case of residual tumors, follow-up continued on a 6-monthly basis. Patients with recurrent or residual tumors are followed closely for the first 2 years and then on an annual basis.

Results

Demographic Data

There were 29 female (64.4%) and 16 male (35.6%) patients, providing a female/male ratio of 1.8:1. The mean age of patients at the time of surgery was 41.7 years (range 16–67). Nineteen tumors (42.2%) were on the right side and 26 (57.8%) on the left side. The radiographic follow-

up period (consisting of serial computed tomography and/or MRI scans) of the series ranged from 6 to 275 months (mean 117.4 months). There were 7 complex cases of TJP combined with vagal paraganglioma on the ipsilateral side and these cases underwent simultaneous excision of both tumors.

Clinical Signs

Hearing loss in 35 patients (77.8%) and tinnitus in 23 cases (51.1%) were the most common presenting complaints. The most common non-otologic symptoms were dysphagia in 23 patients (51.1%), dysphonia in 13 patients (28.9%), and limitation of shoulder movement in 13 patients (28.9%).

Classification and Location of the Tumor

The location and the extent of the lesions were determined preoperatively using high-resolution computed tomography with bone windows and MRI, and were subsequently confirmed intraoperatively. The posterior fossa dura was affected in all cases and other sites of involvement were the cerebellopontine angle (CPA), clivus, internal auditory canal, prepontine cistern, foramen magnum, hypoglossal canal, cavernous sinus, and Meckel's cave (table 1). The tumors were classified according to the Fisch classification [Fisch, 1982] (table 2) with 2 (4.4%) in C1, 20 (44.4%) in C2, 22 (48.9%) in C3 and 1 (2.2%) in C4. There were a total of 27 (60%) cases classified as Di1 and 18 (40%) classified as Di2. In the Di1 group of 27 cases, there were 2 cases (7.4%) in C1, 16 cases (59.2%) in C2, and 9 cases (33.3%) in C3. In the Di2 group of 18 cases, there were 4 cases (22.2%) in C2, 13 cases (72.2%) in C3, and 1 case (5.6%) in C4.

Surgical Procedure

Surgery was performed in a single stage for 29 patients (64.4%), composed of 27 cases in the Di1 group and 2 cases in the Di2 group. Staged surgery was performed in the other 16 patients (35.6%), all of whom were in the Di2 group (table 3). The infratemporal fossa approach type A (IFTA-A) [Fisch, 1988; Sanna, 1995] was the primary approach performed in all 45 cases; however, anterior FN rerouting could not be performed in 5 cases due to FN involvement. For the single-stage procedures, the IFTA-A proved sufficient in 27 cases (93.1%), with 1 case (3.45%) who had an additional labyrinthectomy and 1 case (3.45%) needing an additional transcondylar approach (table 4a). For the staged surgeries, the IFTA-A was the primary approach and the transdural-transsigmoid-transcondylar-transclival [Shin et al., 2011b] approach was performed in 7 cases (43.75%), the modified transcochlear approach [Sanna et al., 1994] in 6 cases (37.5%), with a translabyrinthine, an extreme lateral [Sen and Sekhar, 1990], and a transotic [Fisch, 1988] approach for 1 case (6.25%) each (table 4b). In complex cases with vagal paragangliomas, a transcervical second-stage approach was performed for removal of the vagal component [Sanna et al., 2011; Shin et al., 2011a]. The average

Table 1. Location of the tumor/sites of involvement (total: 45 cases)

	Patients with site involvement
Posterior fossa dura	45 (100.0%)
CPA	7 (15.6%)
Clivus	6 (13.3%)
IAC	3 (6.7%)
Prepontine cistern	3 (6.7%)
Foramen magnum	3 (6.7%)
Hypoglossal canal	2 (4.4%)
Cavernous sinus	2 (4.4%)
Meckel's cave	1 (2.2%)

IAC = Internal auditory canal.

Table 2. Tumor distribution according to the Fisch classification

	C1	C2	C3	C4	Total
Di1	2 (7.4%)	16 (59.2%)	9 (33.3%)	0 (0%)	27 (60.0%)
Di2	0 (0%)	4 (22.2%)	13 (72.2%)	1 (5.6%)	18 (40.0%)
Total	2 (4.4%)	20 (44.4%)	22 (48.9%)	1 (2.2%)	45 (100.0%)

Table 3. Staging of procedures according to the Fisch classification

	Single procedure	Staged procedure
C1Di1	2	–
C2Di1	16	–
C3Di1	9	–
C4Di1	–	–
C1Di2	–	–
C2Di2	–	4
C3Di2	2	11
C4Di2	–	1
Total	29 (64.4%)	16 (35.6%)

Table 4. Surgical approaches used for staged and single-stage procedures

a Single-stage procedures

Procedure	Total
IFTA-A	27 (93.1%)
IFTA-A with TLA	1 (3.4%)
IFTA-A with transcondylar approach	1 (3.4%)
Total	29 (100.0%)

b Staged procedures

1st stage		2nd stage		3rd stage	Total
IFTA-A	+	POTS			7 (43.8%)
IFTA-A	+	TC			6 (37.5%)
IFTA-A	+	TLA			1 (6.2%)
IFTA-A	+	TO			1 (6.2%)
IFTA-A	+	TC	+	EL	1 (6.2%)
Total					16 (100.0%)

TLA = Modified translabyrinthine approach; TC = modified transcochlear approach; TO = transotic approach; POTS = petro-occipital transsigmoid approach.

Table 5. Results of surgical removal according to Fisch classification

	Total removal	Partial removal
C1Di1	2	–
C2Di1	11	5
C3Di1	7	2
C4Di1	–	–
C1Di2	–	–
C2Di2	4	–
C3Di2	6	7
C4Di2	1	–
Total	31 (68.8%)	14 (31.1%)

time interval between the initial and the second procedure was 11 months with a range of 3–21 months. The tumors were frequently found attached to the carotid canal with varying degree of involvement of the ICA, requiring meticulous dissection to achieve complete removal of the tumor. In cases where the intradural component was not excessively large, such as in the majority of Fisch class Di1 cases, approximately 1 cm of the posterior fossa dura was opened and the intradural portion was removed concurrently. The dural opening was then closed primarily with sutures intersewn with a plug of free muscle graft or with strips of abdominal fat inserted into the subarachnoid space.

Preoperative management of the ICA was performed in 16 cases (35.6%), 6–18 weeks before surgery, and consisted of 9 cases of stenting and 7 cases of PBO. PBO had been performed after the demonstration of an adequate collateral supply. The carotid artery was removed in 5 cases of balloon occlusion and 1 case which was a Fisch C4 case with preoperative ICA occlusion by the tumor [Sanna et al., 2004b].

Surgical Outcome

Total tumor removal was achieved in 31 patients (69%), and another 14 patients (31%) underwent a partial resection (table 5). For the cases with total tumor removal, the majority (11 cases, 35.5%) were C2Di1 cases. Resection was performed in a single stage for 20/31 cases (64.5%) and staged for 11/31 cases (35.5%; table 6a). For the cases with partial tumor removal, the majority (5 cases, 35.7%) were C2Di1 cases. Resection was performed in a single stage for 9/14 cases (64.3%) and staged for 5/14 cases (35.7%; table 6b). Recurrence was documented in 3 (9.7%) of 31 cases of total tumor resection, 2 cases of staged surgery, and 1 single-staged procedure, which were managed by a combination of revision surgery with radiotherapy [Sanna et al., 2006] (table 7). The time interval between the first procedure and revision surgery for recurrence ranged from 53 to 136 months. In the 14 cases with partial tumor resection, 7 (50%) residuals were present at the posterior fossa dura, 2 (14.3%) residuals each at the clivus and CPA, and 1 (7.1%) residual each at the cavernous sinus, Meckel's cave, and prevertebral area (table 8a). During the course of follow-up of these residual cases, 8 (57.1%) did not exhibit any growth, while 2 (14.3%) exhibited spontaneous regression. Another 3 (21.4%) were managed with radiation therapy, and 1 (7.1%) underwent revision surgery [Sanna et al., 2006] (table 8b).

CN Status

Thirty-four (75.6%) of the patients were found to be suffering from at least one CN deficit in the preoperative evaluation. The most common CNs involved were the IX and X CN, each involved in 24/34 (70.6%) patients. The IV, V, and VI nerves were also involved each in 1 (2.9%), 1 (2.9%), and 2 (5.9%) patients. The individual preservation rates for the LCNs were as follows: IX (42.8%), X (59%), XI (58%), and XII (64.2%). The overall LCN preservation rate was 56.8% (table 9).

Table 6. Tumor removal for Fisch class C and D tumors

a Distribution of cases with total tumor removal

	Di1				Di2				Total
	C1	C2	C3	C4	C1	C2	C3	C4	
Single	2	11	7	–	–	–	–	–	20 (64.5%)
Staged	–	–	–	–	–	4	6	1	11 (35.5%)
Total	2 (6.5%)	11 (35.5%)	7 (22.6%)	0 (0.0%)	0 (0.0%)	4 (12.9%)	6 (19.3%)	1 (3.2%)	31 (100%)

b Distribution of cases with partial tumor removal

	Di1				Di2				Total
	C1	C2	C3	C4	C1	C2	C3	C4	
Single	–	5	2	–	–	–	2	–	9 (64.3%)
Staged	–	–	–	–	–	–	5	–	5 (35.7%)
Total	0 (0%)	5 (35.7%)	2 (14.3%)	0 (0%)	0 (0%)	0 (0%)	7 (50%)	0 (0%)	14 (100%)

With regard to the FN, 40 patients (88.8%) underwent permanent anterior transposition of the FN. Four patients in this series required a resection of the involved segment of the nerve leaving a defect that was repaired using a sural nerve graft in 3 cases and great auricular nerve in 1 case. Of these 4 cases, 2 patients had reached a HB grade IV and 2 remained in HB grade VI at the end of 1 year. One remaining case underwent a resection of the FN without reconstruction due to complete involvement of the root of the FN. In the group of totally resected cases, 64.5% had a preoperative HB grade I or II status compared to 45% postoperatively, 29% had a preoperative HB grade III status compared to 42% postoperatively, and 6.5% had a HB grade IV–VI status compared to 13% postoperatively. Out of 14 partially resected cases, 42.8% had a preoperative HB grade I or II status compared to 28.6% postoperatively, 28.6% had a preoperative HB grade III status compared to 35.7% postoperatively, and 28.6% had a preoperative HB grade IV–VI status compared to 35.7% postoperatively. Overall, 80% of cases retained a postoperative HB grade I–III status compared to 86.7% preoperatively (table 10, 11).

Perioperative Complications

A single patient developed a HB grade IV FN paresis after embolization. There were no other complications related to embolization. There were 2 (4.4%) cases of CSF leakage in the whole series, and both required an additional procedure for resolution. There was 1 (2.2%) patient who had aspiration pneumonia which resolved

Table 7. Management of recurrence following total removal

	Revision surgery	Combined (surgery and radiation)
Staged	1	1
Single stage	1	0
Total	2 (66.7%)	1 (33.3%)

Table 8. Residual tumors and their management

a Location of residual tumors

	Single stage	Staged	Total
Posterior fossa dura	7	–	7 (50%)
Cavernous sinus	1	–	1 (7.1%)
CPA	1	1	2 (14.3%)
Clivus	–	2	2 (14.3%)
Meckel's cave	–	1	1 (7.1%)
Prevertebral area	–	1	1 (7.1%)
Total	9	5	14 (100%)

b Management of residual tumors

	No growth	Spontaneous regression	Radiation	Revision surgery
Staged	3	0	1	1
Single stage	5	2	2	0
Total	8 (57.1%)	2 (14.3%)	3 (21.4%)	1 (7.1%)

Table 9. LCN preservation rates (total: 45 cases)

	Intact LCNs	
	before operation	after operation
IX	21	9 (42.9%)
X	22	13 (59.1%)
XI	31	18 (58.1%)
XII	28	18 (64.3%)
Overall	102	58 (56.9%)

Table 10. Preoperative FN function according to the HB grading system for cases with partial and total removal

HB grade	Partial removal	Total removal	Total
I or II	6 (42.8%)	20 (64.5%)	26 (57.8%)
III	4 (28.6%)	9 (29%)	13 (28.9%)
IV–VI	4 (28.6%)	2 (6.5%)	6 (13.3%)
Total	14 (31.1%)	31 (68.9%)	45 (100.0%)

Table 11. Postoperative FN function according to the HB grading system for cases with partial and total removal

HB grade	Partial removal	Total removal	Total
I or II	4 (28.6%)	14 (45%)	18 (40%)
III	5 (35.7%)	13 (42%)	18 (40%)
IV–VI	5 (35.7%)	4 (13%)	9 (20%)
Total	14 (31.1%)	31 (68.9%)	45 (100.0%)

spontaneously, and there were no patients who required a tracheostomy. Three patients required delayed phonosurgical procedures for voice restoration after ipsilateral vocal cord paralysis. Two patients with C2Di1 tumors, one after partial removal and another after total removal, underwent a type I thyroplasty, while another patient with a C2De2Di1 tumor that was totally removed had vocal cord medialization with VOX® (Medtronic-Xomed). There were no cases of perioperative meningitis, abscess formation, cerebrovascular accident or mortality in this series. However, 2 patients died during follow-up. One patient died 4 years after the surgery due to unrelated causes and the other died 9 years after the surgery of an unrelated malignancy.

Case Examples

Case 1: Single-Stage Procedure (fig. 1a–c). A 63-year-old woman presented with a 3-month history of dysphonia and mild dysphagia and shoulder weakness noted for 6 months. She had normal hearing function and no family history. She showed normal tympanic membranes on otoscopy and IX, X, and XI CN paralysis. She had a preoperative HB grade I FN status. She was classified as a right-sided class C1Di1 tumor and underwent a single-stage resection. Postoperatively, there was no recovery of the IX, X, and XI CNs, and her FN status postoperatively was HB grade I–II.

Case 2: Staged Surgery (fig. 2a–d). A 67-year-old man presented with dysphagia and facial weakness for almost 7 months. On examination, he had normal otoscopy with a mild right-sided conductive hearing loss as well as a preoperative IX, X, and XI CN status, while his FN status was HB grade III. He was classified as a class C2Di2 tumor, for which an ITFA-A was performed as the first stage. Following successful resection of the extradural component, he underwent resection of the intradural component using a transdural-transsigmoid-transcondylar-transclival access. The tumor was completely resected and, postoperatively, he had palsies of the IX, X, and XI nerves, and his postoperative FN status immediately after surgery was HB grade V, but he has subsequently recovered to a grade IV HB.

Discussion

TJPs typically expand into the subarachnoid space by penetrating the dura of the posterior fossa along the LCNs. Less commonly, they can also invade intradurally through the internal auditory canal and the dura of the middle fossa [Jackson et al., 1992; Sanna et al., 2004a]. Our data also supports this theory, and some of the cases with extension into the CPA and cavernous sinus might have had direct invasion through the petrous bone. The majority of Di1 cases were C2 (59.2%) and Di2 cases were C3 (72.2%), which shows that the degree of intradural extension, intracranial and carotid involvement correlates well with the increasing class of tumor as reported previously [Patel et al., 1994; Sanna et al., 2004a].

Rationale for Staging

Even though the ideal primary treatment for TJPs is total surgical extirpation [Jackson et al., 1996], total resection of an intradural extension is challenging due to the comparative inaccessibility of these extremely vascu-

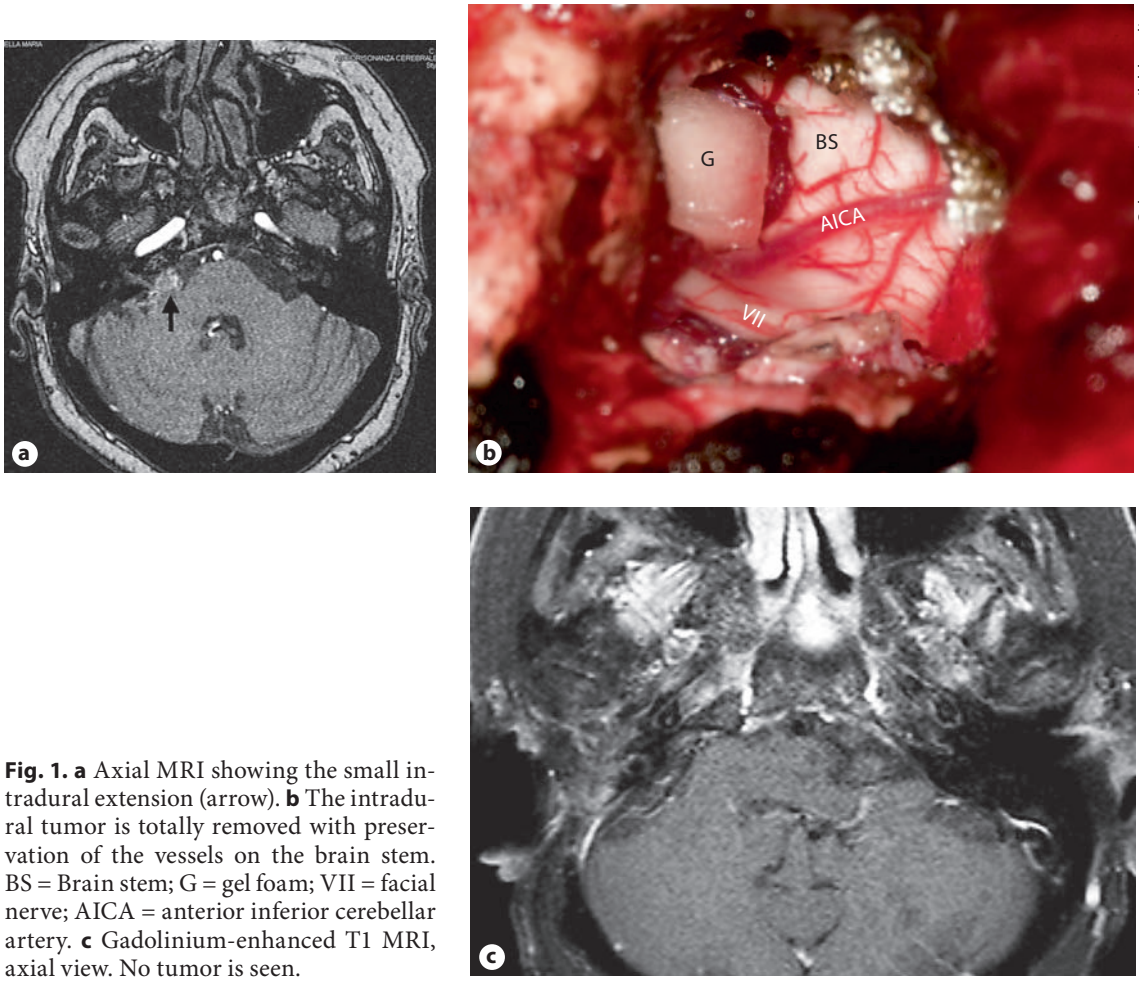

Fig. 1. a Axial MRI showing the small intradural extension (arrow). **b** The intradural tumor is totally removed with preservation of the vessels on the brain stem. BS = Brain stem; G = gel foam; VII = facial nerve; AICA = anterior inferior cerebellar artery. **c** Gadolinium-enhanced T1 MRI, axial view. No tumor is seen.

lar tumors nestling among a cluster of vital structures. In the presence of an intradural component, there are conflicting opinions in the literature regarding the criteria for staged surgery [Kinney, 1980; Anand et al., 1993; Gjuric et al., 1996; Jackson et al., 2001]. We prefer a staged procedure for tumors with more than 2 cm intradural extension to prevent postoperative CSF leaks, and one-stage removal for tumors with an intradural extension of ≤2 cm [Fisch et al., 1984; Sanna, 2008]. While the reported rate of postoperative CSF leaks in surgery for intracranial extension and complex cases ranged from 14.2 to 33.3% [Netterville and Civantos, 1993; Patel et al., 1994; Al-Mefty and Teixeira, 2002; Magliulo et al., 2008], it occurred in only 2 out of 45 cases (4.4%) in our study. Lumbar drains are not routinely used in our center. Staged surgery avoids direct connection between the subarachnoid space and large spaces of the neck avoiding CSF leaks and related complications. When performing a single-stage resection, concerns regarding the creation of a large skull base and dural defect can create problems. Wide bone removal with a large dural opening, essential to expose the tumor and minimize recurrence, may be curtailed out of concern for postoperative CSF leaks, which can impede safe removal of intradural tumors. A distinct advantage that we have observed over the years has been the relative ease and technical simplicity of the intradural tumor removal in the second stage. Because the remaining tumor has frequently been devascularized after the initial stage, dissection and preservation of the LCNs in the second intradural stage have been simplified to a great degree, which is further aided by the embolization of the blood supply before the second stage to further reduce intraoperative bleeding. While repeat angiography and embolization are disadvantages of a staged procedure, we are convinced that the benefits of the embolization outweigh the risks of neurological complications

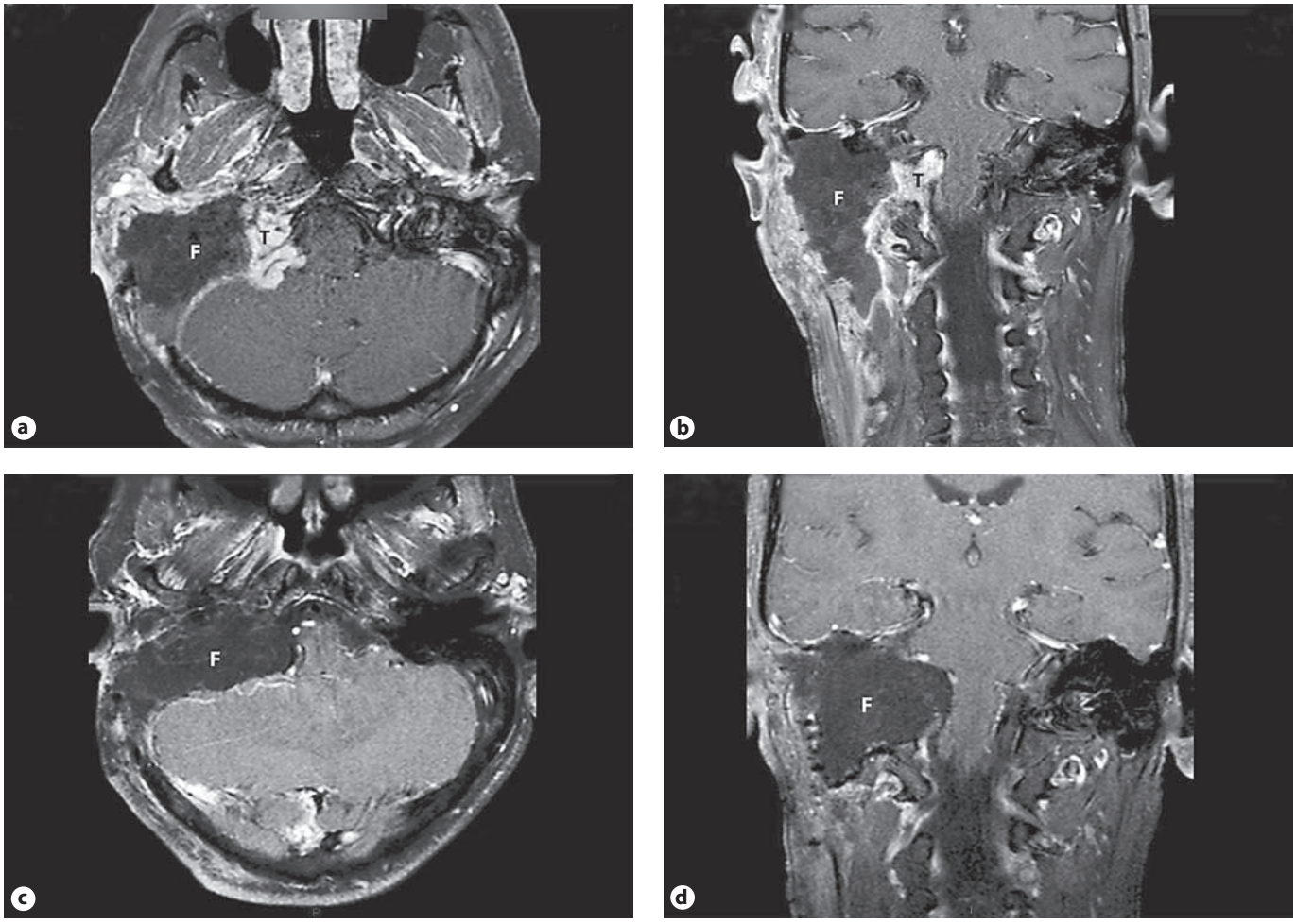

Fig. 2. a, **b** MRI, axial and coronal views after the first-stage surgery. The residual intradural tumor is noted. The surgical defect is filled with abdominal fat. T = Intradural tumor; F = fat. **c**, **d** MRI, axial and coronal views after the second-stage surgery. After the surgery, there is no residual tumor.

as we have noted only 1 patient having a HB grade IV FN paresis after embolization. In order to permit repeated embolization, utmost care is placed on surgical manipulation of the external carotid artery during first-stage resection.

Another problem with single-stage surgeries for tumors with large intradural extensions is an issue of surgeon fatigue that is often overlooked. These complex cases are some of the most challenging for a skull base surgeon and can be expected to continue for >8 h. As the intracranial component is removed following extradural resection in single-stage operation, the most critical and delicate dissection is carried out when fatigue is most likely.

We believe the advanced level of extension and carotid involvement in this series warranted the use of the IFTA-A as primary procedure in the majority of the cases. Infiltrated bone around the jugular fossa cannot be safely and adequately removed without rerouting the FN, particularly if there is anterior extension along the carotid canal as with the majority of our cases [Oghalai et al., 2004; Sanna et al., 2004a, 2004b; Shin et al., 2011b]. In addition, when performing the IFTA-A, the FN should be anteriorly transposed permanently. This minimizes the risk to the FN if subsequent surgery is required. It is important, however, to leave some constant bony landmarks to allow orientation for the second-stage surgery. Even if a translabyrinthine approach is required in pri-

mary surgery, conservation of a portion of the labyrinth is important. Small areas of infiltrated dura can be easily resected and repaired. If an extensive area is of concern, a second-stage resection can be considered. During the second stage, the approach is determined by the location and size of the residual tumor and the patient's hearing function. The modified transcochlear approach [Sanna, 2008] and a transdural-transsigmoid-transcondylar-transclival approach [Shin et al., 2011b] were preferred in most cases, which provided better access from the jugular foramen to the CPA and the LCN [Sanna et al., 1994; Shin et al., 2011b]. Other approaches that can be utilized are the translabyrinthine, an extreme lateral or transotic approach. Concerning the order of resection, our practice is that both in single and staged procedures, the resection of the jugular fossa and extradural components should precede dural opening and resection. We have found that there is a significant reduction in the vascularity of the intradural tumor following extradural resection, which facilitates removal. Additional vascularity is usually drawn from the posterior inferior cerebellar artery, which must be identified at the time of resection. Bipolar coagulation of the tumor surface is used extensively prior to debulking and removal. Encasement of the posterior inferior cerebellar artery, anterior inferior cerebellar artery, extension to the cavernous sinus, and brain parenchymal invasion are all indications for sub-total removal.

Skull Base Reconstruction

The main aim in the closure of skull base defects is to prevent the occurrence of CSF leak. Cosmetic and functional considerations also play a significant role. During the initial soft-tissue approach, preservation of the multiple fascial layers is essential to facilitate a watertight closure. The skin is raised above the temporoparietal fascia, allowing the use of this flap if required [Kaylie et al., 2007]. However, we do not routinely utilize the temporalis flap for closure. The temporalis fascia and periosteum must be incised in a fashion that allows primary closure. Transection of the external auditory canal results in a degree of tissue loss, and when the strong periosteum and soft tissue of the tympanic bone are removed during an IFTA-A, maintenance of firm pressure on the fat graft is difficult, significantly increasing the risk of CSF leak if the dura has been opened. In order to avoid this problem, we close the external auditory canal in a cul-de-sac manner. The external auditory canal with tragal cartilage is then sutured to the subcutaneous tissue.

Obliteration of dead space is an integral part of closure in these defects. Muscle and fat provide a malleable tissue that can be packed into complex cavities, which facilitates augmentation of wound and dural closure. Infection of the fat graft is not a problem in this setting and, indeed, it has been reported to have inherent immunoreactive properties [Fantuzzi, 2005]. It provides bulk to reconstructed deficits improving cosmesis, is used to primarily close small dural defects, and provides a contrast medium for follow-up MRI scans.

The temporalis and sternocleidomastoid muscles are widely used as local muscle flaps [Ramina et al., 2005]. However, both flaps have a limited arc of rotation, can atrophy, and are associated with cosmetic defects [Neligan et al., 1996]. Therefore, they should not be relied upon to prevent CSF leak with a large dural defect. We use a simple and robust musculofascial closure by suturing the previously detached sternocleidomastoid muscle to the temporalis fascia facilitating the secure placement of the abdominal fat graft.

While we advocate that any significant dissection of the neck with a coexisting large dural defect be avoided, small dural defects, usually through the intracranial opening of the jugular fossa, can be closed with strips of abdominal fat, augmented with fibrin glue. This is followed by the routine placement of abdominal fat to obliterate the remainder of the defect and multilayer myofascial closure with 2-layer skin closure.

There is an increasing trend toward single-stage resection of intradural and extradural disease [Neligan et al., 1996; Gullane et al., 2005]. This is certainly due to improved reconstruction techniques, especially in the area of free flap reconstruction. Despite these improvements, the rate of postoperative CSF leaks remains significant [Jackson et al., 2004]. We feel that in circumstances where these techniques are required, the patient is better served by a staged procedure, avoiding the risks of free tissue transfer. In addition, it avoids the perioperative use of lumbar drainage, employed by most units performing single-stage resections [Kaylie et al., 2007].

Surgical limitation

In each of the 14 residual cases, all possible outcomes had been extensively discussed with the individual patient and a final decision was made in line with their wishes. In 2 patients, small tumor remnants in the cavernous sinus were deliberately left behind to avoid compromising CNs III, IV, and VI. A dominant sigmoid sinus, basilar and vertebral artery (VA) involvement, absence of collateral flow on temporary occlusion of

Table 12. Comparison of outcome measures among previously published data and present series

Reported series	Patients with intradural extension	Staged or single procedure	Main surgical approach	Gross total resection	Perioperative CSF leaks
Patel et al., 1994	10	Staged	1st: Transtemporal approach with a preauricular subtemporal approach 2nd: Retrosigmoid and extreme lateral transcondylar approaches	8 (80%)	4 (40 %)
Jackson et al., 2004	22	Single	Not described	20 (89%)	3 (6%)
Magliulo et al., 2008	11	Staged	1st: Infratemporal fossa approach 2nd: Retrosigmoid approach	8 (72%)	2 (18%)
Moe et al., 1999	58	Single mainly Di1 Staged mainly Di2–3	1st: IFTA-A and -B 2nd: Neurosurgical procedure	36 (62%)	6 (11%)
Present study	45	Single mainly Di1 Staged mainly Di2–3	1st: IFTA-A 2nd: TD-TS-TC-TCl approach/transcochlear/transotic/extreme lateral/translabyrinthine	31 (69%)	2 (4%)

TD-TS-TC-TCl = Transdural-transigmoid-transcondylar-transclival.

the ICA, and advanced age with a poor general condition were other factors that necessitated a partial resection. We believe the small, often miniscule residual tumor is best managed with a wait and watch policy with serial MRI scans. Our data also revealed that residual components showed no growth in 8 patients (57.1%) and regressed in 2 patients (14.3%). If growth can be demonstrated, which is uncommon as a result of devascularization of the remnant, additional surgery or radiosurgery is a viable option in such a scenario [Sanna et al., 2006].

LCN Preservation

Regarding preservation of preoperatively functioning LCNs in the cases with intradural extension, the proximity of these structures to an infiltrated medial wall of the jugular bulb makes it a disappointing endeavor, as infiltration of the nerves can occur despite normal preoperative function [Sen et al., 2001]. Thus, if the goal is total removal, invasion of the medial wall of the jugular bulb by tumors necessitates its resection with a correspondingly high risk for CN deficits [Jackson et al., 2004; Oghalai et al., 2004]. Otherwise, the alternative in the setting of preoperatively functioning LCNs is to leave residual tumor surrounding these nerves [Oghalai et al., 2004]. However, this can compromise the integrity of the resection, especially in younger patients who may be better served by a more aggressive resection in light of the fact that they tend to compensate better than older patients [Oghalai et al., 2004]. Moreover, age and preoperative medical fitness are all factors to be taken into consideration in making these decisions, and the presence of contralateral disease with a risk of bilateral LCN paralysis changes this aggressive approach with serial imaging and observation more likely in these situations [Jackson et al., 2004; Oghalai et al., 2004]. In our series, in spite of the extensive nature of these intradurally extending tumors, we managed to maintain a 56.8% overall preservation rate of LCN (table 8) with a correspondingly low incidence of perioperative deglutition and voice complications.

Literature Review

In order to compare our results, we conducted a literature review on PubMed, concentrating on series dealing with intradural extension of TJPs (table 12), with gross total tumor resection and rate of CSF leak as the comparative outcome measures. Jackson et al. [2004] utilized a single-stage surgery for 22 cases. They did not specify a specific approach in their paper, with gross total resection in 89% of cases and a 6% rate of CSF leak. Moe et al. [1999] did not clearly specify in their paper

Table 13. Proposed revision to the Fisch classification for tympanic and tympanojugular paragangliomas

Class A	Tumors limited to the middle ear
A1	Tumors completely visible on otoscopic examination
A2	Tumor margins are not visible on otoscopy; tumor may extend anteriorly to the Eustachian tube and/or to the posterior mesotympanum
Class B	Tumors limited to the tympanomastoid area without destruction of bone in the infralabyrinthine compartment of the temporal bone
B1	Tumors confined to the middle ear cleft with extension to the hypotympanum
B2	Tumors involving the middle ear cleft with extension to the hypotympanum and the mastoid
B3	Tumors confined to the tympanomastoid compartment with erosion of the carotid canal
Class C	Tumors extending, destroying bone of the infralabyrinthine and apical compartment of the temporal bone and involving the carotid canal
C1	Tumors with limited involvement of the vertical portion of the carotid canal
C2	Tumors invading the vertical portion of the carotid canal
C3	Tumors with invasion of the horizontal portion of the carotid canal
C4	Tumors reaching the anterior foramen lacerum
Class D*	Tumors with intracranial extension
Di1	Tumors up to 2 cm intradural extension
Di2	Tumors with more than 2 cm intradural extension
Di3	Tumors with inoperable intradural extension
Class V	Tumors involving the vertebral artery
Ve	Tumors involving the extradural vertebral artery
Vi	Tumors involving the intradural vertebral artery

* Denotes new proposed class D classification with removal of De component.

whether or not staged surgery was used, but we have assumed that they utilized a staged procedure for Di2 cases, with the intradural portion removed at a second-stage neurosurgical operation as specified by Fisch [1988] previously. They utilized a combination of ITFA type A and B approaches for tumors with intradural extension achieving gross total resection in 62% of the cases with an 11% rate of CSF leak. However, of the 23 cases with Di2 extension in their study, 14 (61%) had a subtotal excision. Patel et al. [1994] and Magliulo et al. [2008] advocated a staged approach to these tumors. Patel et al. [1994] utilized a first-stage transtemporal and preauricular subtemporal approach with a second-stage retrosigmoid/extreme lateral/transcondylar approach for 10 cases, with gross total resection in 80% of cases and a 40% rate of CSF leak. Magliulo et al. [2008] utilized a first-stage ITFA-A with a second-stage retrosigmoid approach for 11 cases, with gross total resection in 72% of cases and an 18% rate of CSF leak. In our present series of 45 cases, we have achieved gross total tumor resection in 69% of cases with a 4.4% rate of CSF leak, which compares very favorably with the previously published data and, in our opinion, provides a compelling argument for the rationale of staging the procedure for cases with >2 cm of intradural extension.

Proposed Revision of the Fisch Classification

We have previously recommended a small addition to the Fisch classification to reflect the involvement of the VA [Shin et al., 2011b]. We suggested adding two adjunct classes regarding the VA involvement in order to correctly plan the surgical approach. Class Ve refers to TJP involving the extradural tract of the VA going from the transverse foramen of C6 to C1 (foraminal or V2) and through the transverse foramen of C1 to the foramen magnum (extraspinal or V3). Class VI regards involvement of the V4 tract of the VA (going from the foramen magnum to the basilar artery) that in our experience is inevitably associated with a large intradural extension. In line with our experience in dealing with these large intradural TJPs, and in concert with the data from this present study, we have realized that the Fisch classification can be further streamlined. We have noted that for the class D tumors, the tumor has already penetrated the cranial cav-

ity in both De and Di components and the general surgical approach for excision utilized for both De and Di components is almost the same [Sekhar and Oliveira, 1999; Sanna et al., 2004a; Sanna, 2008]. However, once the tumor extends intracranially, the presence of the Di subcomponent is the more important criterion that influences the details of the approach that should be used and if staged surgery is necessary. The De subcomponent has less of an influence on the prospective management as compared to the Di subcomponent. Therefore, in our opinion, in line with the constantly evolving methods and techniques of skull base surgery, we recommend that the De subcomponent for class D tumors be removed evolving the Fisch classification to better reflect the relative clinical merits of its various subcomponents (table 13). The revised classification for class D tumors would be a simpler and more accurate reflection of the actual practices in many skull base centers, including ours, where the main criteria in determining the management of these unfortunate patients is the degree of intradural extension.

Conclusion

Management of TJP with intradural extensions are challenging endeavors and maximizing the surgical outcome while minimizing morbidity is ultimately what every surgeon aims for. In our study of these 45 patients, total tumor removal was achieved in 69% of cases, the rate of postoperative CSF leakage was 4.4%, and overall preservation rate of LCN was 56.8%. We believe, in the presence of a large intradural extension, the practice of staged surgery with sophisticated preoperative intervention and intraoperative techniques is the principal method for successful tumor resection with an extremely low level of postoperative complications.

Acknowledgement

This study was supported by a grant from the Associazione Italiana Neuro-Otologica.

Disclosure Statement

The authors have no conflicts of interest to disclose.

References

Al-Mefty O, Teixeira A: Complex tumors of the glomus jugulare: criteria, treatment, and outcome. J Neurosurg 2002;97:1356–1366.

Anand VK, Leonetti JP, Al-Mefty O: Neurovascular considerations in surgery of glomus tumors with intracranial extensions. Laryngoscope 1993;103:722–728.

Fantuzzi G: Adipose tissue, adipokines, and inflammation. J Allergy Clin Immunol 2005; 115:911–919; quiz 920.

Fisch U: Infratemporal fossa approach for glomus tumors of the temporal bone. Ann Otol Rhinol Laryngol 1982;91:474–479.

Fisch U, Fagan P, Valavanis A: The infratemporal fossa approach for the lateral skull base. Otolaryngol Clin North Am 1984;17:513–552.

Fisch U: Microsurgery of the Skull Base. Stuttgart, Thieme, 1988.

Gjuric M, Rudiger Wolf S, Wigand ME, Weidenbecher M: Cranial nerve and hearing function after combined-approach surgery for glomus jugulare tumors. Ann Otol Rhinol Laryngol 1996;105:949–954.

Gullane PJ, Lipa JE, Novak CB, Neligan PC: Reconstruction of skull base defects. Clin Plast Surg 2005;32:391–399, vii.

House JW, Brackmann DE: Facial nerve grading system. Otolaryngol Head Neck Surg 1985; 93:146–147.

Jackson CG, Haynes DS, Walker PA, Glasscock ME 3rd, Storper IS, Josey AF: Hearing conservation in surgery for glomus jugulare tumors. Am J Otol 1996;17:425–437.

Jackson CG, Kaylie DM, Coppit G, Gardner EK: Glomus jugulare tumors with intracranial extension. Neurosurg Focus 2004;17:E7.

Jackson CG, McGrew BM, Forest JA, Netterville JL, Hampf CF, Glasscock ME 3rd: Lateral skull base surgery for glomus tumors: long-term control. Otol Neurotol 2001;22:377–382.

Jackson CG, Netterville JL, Glasscock ME 3rd, Hampf CR, Carrasco VN, Haynes DS, Strasnick B, Fisher J: Defect reconstruction and cerebrospinal fluid management in neurotologic skull base tumors with intracranial extension. Laryngoscope 1992;102:1205–1214.

Kaylie DM, O'Malley M, Aulino JM, Jackson CG: Neurotologic surgery for glomus tumors. Otolaryngol Clin North Am 2007;40:625–649, x.

Kinney SE: Glomus jugulare tumor surgery with intracranial extension. Otolaryngol Head Neck Surg 1980;88:531–535.

Magliulo G, Parrotto D, Alansi W, Cuiuli G, Alla FR: Intradural jugular paragangliomas: complications and sequelae. Skull Base 2008; 18:189–194.

Moe KS, Li D, Linder TE, Schmid S, Fisch U: An update on the surgical treatment of temporal bone paraganglioma. Skull Base Surg 1999;9: 185–194.

Neligan PC, Mulholland S, Irish J, Gullane PJ, Boyd JB, Gentili F, Brown D, Freeman J: Flap selection in cranial base reconstruction. Plast Reconstr Surg 1996;98:1159–1166; discussion 1167–1168.

Netterville JL, Civantos FJ: Defect reconstruction following neurotologic skull base surgery. Laryngoscope 1993;103:55–63.

Oghalai JS, Leung MK, Jackler RK, McDermott MW: Transjugular craniotomy for the management of jugular foramen tumors with intracranial extension. Otol Neurotol 2004;25: 570–579; discussion 579.

Patel SJ, Sekhar LN, Cass SP, Hirsch BE: Combined approaches for resection of extensive glomus jugulare tumors. A review of 12 cases. J Neurosurg 1994;80:1026–1038.

Ramina R, Maniglia JJ, Fernandes YB, Paschoal JR, Pfeilsticker LN, Coelho Neto M: Tumors of the jugular foramen: diagnosis and management. Neurosurgery 2005;57:59–68; discussion 59–68.
Sanna M: Atlas of Temporal Bone and Lateral Skull Base Surgery. Stuttgart, New York, Thieme, 1995.
Sanna M: Atlas of Microsurgery of the Lateral Skull Base, ed 2. Stuttgart, New York, Thieme, 2008.
Sanna M, De Donato G, Piazza P, Falcioni M: Revision glomus tumor surgery. Otolaryngol Clin North Am 2006;39:763–782, vii.
Sanna M, Jain Y, De Donato G, Rohit, Lauda L, Taibah A: Management of jugular paragangliomas: the Gruppo Otologico experience. Otol Neurotol 2004a;25:797–804.
Sanna M, Mazzoni A, Saleh EA, Taibah AK, Russo A: Lateral approaches to the median skull base through the petrous bone: the system of the modified transcochlear approach. J Laryngol Otol 1994;108:1036–1044.
Sanna M, Piazza P, Ditrapani G, Agarwal M: Management of the internal carotid artery in tumors of the lateral skull base: preoperative permanent balloon occlusion without reconstruction. Otol Neurotol 2004b;25:998–1005.
Sanna M, Shin SH, De Donato G, Sivalingam S, Lauda L, Vitullo F, Piazza P: Management of complex tympanojugular paragangliomas including endovascular intervention. Laryngoscope 2011;121:1372–1382.
Sekhar LN, Oliveira ED: Cranial Microsurgery: Approaches and Techniques. New York, Thieme, 1999.
Sen C, Hague K, Kacchara R, Jenkins A, Das S, Catalano P: Jugular foramen: microscopic anatomic features and implications for neural preservation with reference to glomus tumors involving the temporal bone. Neurosurgery 2001;48:838–847; discussion 847–848.
Sen CN, Sekhar LN: An extreme lateral approach to intradural lesions of the cervical spine and foramen magnum. Neurosurgery 1990;27:197–204.
Shin SH, Piazza P, De Donato G, Sivalingam S, Lauda L, Vitullo F, Sanna M: Management of vagal paragangliomas including application of internal carotid artery stenting. Audiol Neurotol 2011a;17:39–53.
Shin SH, Sivalingam S, De Donato G, Falcioni M, Piazza P, Sanna M: Vertebral artery involvement by tympanojugular paragangliomas: management and outcomes with a proposed addition to the Fisch classification. Audiol Neurotol 2011b;17:92–104.