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An Update on the Surgical Treatment of Temporal Bone Paraganglioma

ABSTRACT—In 1982, Fisch described his results for the surgical treatment of 74 paragangliomas of the temporal bone, 5 years after his description of the infratemporal fossa approaches (types A and B). This study reviews the subsequent experience of the Department of Otolaryngology—Head and Neck Surgery of the University of Zürich with more than 136 surgically treated cases of paraganglioma of the temporal bone and discusses our current therapy 20 years after the initial description. One hundred nineteen (90%) of the patients had advanced tumors (Fisch class C or C+D), and 81 (68%) had intracranial extension. Total tumor excision was possible in 109 (82%) patients. Subtotal excision was performed in 22 (17%) patients, 21 of whom had intradural tumor invasion. In these cases, the resection was limited not by actual tumor size but by the degree of intracranial intradural tumor extension. Partial tumor excision was undertaken in only 1 patient with a C4De2Di2 tumor. The success rate in preservation of function of the lower cranial nerves was encouraging. Of the 69 patients whose facial nerve status was followed postoperatively, 81% maintained Fisch grade 76 to 100% (House-Brackman grades I and II). Analysis of follow-up data ranging from 2 to 11 years demonstrated 98% disease-free survival when total tumor extirpation was possible. In the patients who underwent subtotal or partial surgical resection there has been no subsequent tumor growth detected by either clinical or neuroradiological evaluation. We have confirmed after more than 20 years of experience that the infratemporal fossa approaches are a safe, highly effective means of surgical management of paragangliomas of the temporal bone, allowing eradication or arrest of disease with minimal morbidity. Limited intradural surgical resection in cases of very extensive tumors can greatly benefit patients for whom complete excision is not an option.

The management of paraganglioma of the temporal bone is uniquely challenging for a number of reasons. The area in which they arise is anatomically complex and involves critical neurovascular structures. Although they are histologically benign they may behave locally as if malignant, with extensive invasion of bone, soft tissue, and nerves.[1] They may be solitary or multicentric, sporadic or familial. Because patients often intially present with mild symptoms and the course of the disease may be relatively innocuous, the diagnosis is com-

Skull Base Surgery, Volume 9, Number 3, 1999. Department of Otolaryngology—Head and Neck Surgery, University of Zürich Hospital, Zürich, Switzerland (KM, TL, SS, UF), and Department of Otolaryngology—Head and Neck Surgery, The Johns Hopkins University, Baltimore, Maryland (DL). Reprint requests: Dr. U. Fisch, Department of Otolaryngology—Head and Neck Surgery, University Hospital of Zürich, Frauenklinikstr. 24, 8091 Zürich, Switzerland. 1052–1453/1999/E1098–9072(1999)09:03:0185–0194:SBS00148X

monly missed for years. In addition, the literature on this topic is confusing and misleading, with considerable variation in aspects as fundamental as tumor terminology and classification. Thus it is not surprising that the treatment of paragangliomas has not been standardized, and debate still rages on whether surgery, radiation, or simply observation is best.

Our current treatment philosophy has been developed and modified by research and experience over three decades. During this time, advances in diagnostic imaging and interventional neuroradiology allowed improved evaluation of tumor extent and the involvement of adjacent structures. This made the development of a classification system to aid in the description and discussion of temporal bone paragangliomas possible.[2] In 1977, the group of infratemporal fossa approaches for the surgical management of these tumors was outlined,[3] with permanent anterior rerouting of the facial nerve and exposure of the intratemporal course of the internal carotid artery (ICA).[3] These approaches allowed complete removal of even highly advanced lesions, including those situated within the infralabyrinthine and apical compartments of the temporal bone and have become the standard route for excision of these tumors, with significant reduction in morbidity and mortality.[4,5]

The purpose of this study is therefore to present the evolution of our treatment to date, presenting our recent experience and current tumor management protocol based on the review of 136 patients treated for paragangliomas of the temporal bone between 1983 and 1994.

MATERIALS AND METHODS

Patient Evaluation

The medical records of 136 patients who underwent surgical removal of temporal bone paragangliomas from January 1983 to December 1994 in the Department of Otolaryngology—Head and Neck Surgery, University of Zürich Hospital, were reviewed. Four patients with incomplete documentation could not be included in all aspects of the study.

All patients had undergone extensive preoperative and postoperative evaluations. The complete preoperative investigation consisted of a detailed case history, a thorough head and neck examination with particular attention to neurological deficits, audiometry, and vestibular testing. Radiological evaluation included preoperative and postoperative high-resolution computed tomography (HRCT) or magnetic resonance imaging (MRI), or both, along with selective angiography. Tumors were classified according to the Fisch classification (Table 1).

Preoperative facial nerve function was available in the case history; the Fisch grading system[6] was used in all instances (Table 2; for conversion to House-Brackman grading system, see Table 12). Sixty-nine patients were available for postoperative facial nerve analysis either directly, or via review of medical photography.

Surgical Approaches

Details of the surgical steps for the procedures used have been described previously.[7] Since 1986 intraoperative monitoring of the seventh, tenth, and twelfth cranial nerves was used in all cases. The approaches employed by class of paraganglioma were the following:

Class A: transcanal, transmastoid
Class B: transmastoid, canal wall-up with temporary resection and reconstruction of the canal wall; subtotal petrosectomy
Class C1: infratemporal fossa approach type A
Class C2: infratemporal fossa approach type A
Class C3: infratemporal fossa approach type A
Class C4: infratemporal fossa approach type A and type B.

Table 1. Fisch Classification of Temporal Bone Paragangliomas

Class A (glomus tympanicum)	Limited to mesotympanum
Class B (glomus hypotympanicum)	Limited to hypotympanum, mesotympanum, and mastoid without erosion of jugular bulb
Class C	Involvement and destruction of infralabyrinthine and apical compartments. Subclassification by degree of carotid canal erosion
C1	No invasion of carotid; destruction of jugular bulb/foramen
C2	Invasion of vertical carotid canal between foramen and bend
C3	Invasion along horizontal carotid canal
C4	Invasion of foramen lacerum and along carotid into cavernous sinus
Class D	Intracranial extension (De, extradural; Di, intradural)
De1	Up to 2-cm dural displacement
De2	More than 2-cm dural displacement
Di1	Up to 2-cm intradural extension
Di2	More than 2-cm intradural extension

Table 2. Fisch Facial Nerve Grading System*

Facial Symmetry	*%*	*Points*
At rest	0	0
	30	6
	70	14
	100	20
Wrinkling forehead	0	0
	30	3
	70	7
	100	10
Closing eyes	0	0
	30	9
	70	21
	100	30
Smiling	0	0
	30	9
	70	21
	100	30
Whistling	0	0
	30	3
	70	7
	100	10

*Points are totaled to express overall facial function as a percentage.
0% = complete asymmetry with no voluntary movement; 30% = poor symmetry; 70% = fair symmetry; 100% = complete symmetry

Evaluation of Tumor Removal

The evaluation of extent of tumor removal was made by review of both surgical reports and postoperative HRCTs and MRIs that were routinely obtained within 3 months after surgery. The following terms were used to describe the tumor removal:

1. Total resection: *no residual tumor*
2. Subtotal resection: *more than 95% of tumor removed*
3. Partial resection: *more than 50% of tumor removed*
4. Enlarged biopsy: *less than 50% of tumor removed*

RESULTS

Patient Population

The patients ranged in age from 14 to 75 years, with a mean age of 44 years. Forty-six (35%) of the patients were men, and 86 (65%) were women. There was equal prevalence of left-sided (52%) and right-sided (48%) lesions. Eighteen patients (14%) had multiple paragangliomas for a total of 47 tumors (unilateral or bilateral). Seven patients (5%) had a family history of similar tumors. Multiple paragangliomas occurred in 3 (43%) of these patients, who ranged in age from 21 to 47 years, with a mean of 32 years.

Twenty-six (20%) patients had previously undergone surgery for removal of the paraganglioma. Twelve of these 26 had had more than two operations, and 1 patient had had 6 prior attempts at extirpation.

Presenting Symptoms

The most common initial symptom was pulsatile tinnitus, which occurred alone in 31 (24%) patients or in combination with hearing loss in 36 (28%) patients for a total of 67 (52%) patients (Table 3). The most common symptoms at the time of diagnosis were hearing loss and pulsatile tinnitus, which were noted in 112 (85%) and 110 (83%) of patients, respectively (Table 4). The mean interval between the onset of these symptoms and diagnosis was 2.6 years (0 to 23 years). Five patients (4%) were found to have catecholamine-secreting paragangliomas.

Tumor Characteristics

Only 10% of the patients in this series had limited (class A or B) tumors. Ninety percent had advanced (class C or C + D) tumors, 72% of which extended intracranially. Table 5 shows the distribution of tumors by

Table 3. Initial Symptoms in Paraganglioma of the Temporal Bone

Symptom	*Number of Patients*	*%*
Hearing loss and pulsatile tinnitus	36	28
Pulsatile tinnitus (alone)	31	24
Facial weakness	19	15
Hearing loss	13	10
Aural pain	7	5
Hoarseness	6	5
Aural pressure	5	4
Dysphagia	5	4
Aural bleeding	3	2
Vertigo	1	1
None	2	2
Total	128	100

Table 4. Clinical Findings at Time of Presentation (N = 132)

Symptoms	*Number of Patients*	*%*
Pulsatile tinnitus	110	83
Hoarseness	36	27
Vertigo	32	24
Dizziness	29	22
Dysphagia	25	19
Aural pressure	15	11
Otalgia	27	20
Headache	8	6
Cough	3	2
Signs	*Number of Patients*	*%*
Hearing loss	112	85
Conductive	52	
Sensorineural	1	
Mixed	59	
Middle ear mass	89	68
Partial facial palsy	41	32
External auditory canal mass	33	25
Aural bleeding	14	11
Total facial palsy	9	7

Table 6. Distribution of Class C Temporal Bone Paragangliomas by Subclassification

Subclass	*Number of Patients*	*%*
C1	11	12
C1De1	3	
C2	16	33
C2De1	15	
C2De1Di1	7	
C2De2Di1	2	
C3	5	38
C3De1	7	
C3De1Di1	7	
C3De1Di2	2	
C3De2	2	
C3De2Di1	15	
C3De2Di2	7	
C4	1	17
C4De1	1	
C4De1Di1	1	
C4De2Di1	3	
C4De2Di2	14	
Total	119	100

stage, and Table 6 demonstrates the classification of the advanced tumors into C and D subsets.

There were no cranial nerve deficits in patients with class A and B tumors (Table 7). Conversely, in patients with class C and D tumors, the severity of cranial nerve involvement was proportional to the size of the mass. The most common cranial nerve involved was the cochlear nerve (mixed hearing loss, 48%), followed by the tenth (40%), seventh (39%), ninth (dysphagia, 34%), twelfth (31%), and sixth (5%) cranial nerves. Of those with a preoperative facial nerve deficit, 9 of 52 (17%) had complete facial paralysis.

Surgical Treatment

Preoperative embolization was performed for 114 (90%) of the patients. Preoperative balloon test occlusion of the ICA was performed in 35 (29%) patients who had C3 or C4 tumors. Table 8 summarizes the surgical approaches used for each class of tumor. All of the 8 class A tumors were removed via either a transcanal or a transmastoid approach. Excision of the 5 class B tumors was performed by a transmastoid approach in 2 patients, subtotal petrosectomy in 2 patients, and infratemporal fossa type A in 1 patient. Of the 14 class C1 tumors, 2 were removed by a transmastoid approach and 2 by subtotal petrosectomy, and 10 required the infratemporal approach type A. Only 1 of the C2 tumors could be excised by subtotal petrosectomy; the other 39 were accessed by the type A approach. All 45 of the C3 tumors required the type A approach. The 20 C4 tumors (19 with D extension) in this series were extirpated by the type A approach in 16 cases (80%) and by combined type A and B approaches in 4 instances (20%) (all with D extension).

Thus, the transcanal approach can be used only for class A tumors (2% of all cases), the transmastoid approach for class A and B and limited C1 tumors (7% of all cases), and the subtotal petrosectomy for larger class B, C1, and limited C2 cases (4% of all cases). The infratemporal fossa type A approach was used for an advanced class B tumor and class C1 to C4 tumors (84% of all cases). A combined type A + B approach was required in 3% of the patients, all of whom were class C4.

The mean operative time for all tumor classes was 6 hours and 5 minutes (range 25 minutes to 9 hours, 30 minutes) and was related to tumor extent. The estimated blood loss was likewise related to tumor class and averaged 2.0 L (range 0 to 10.6 L). As expected, patients with larger tumors also required longer hospitalization, the average having been 18 days (Table 9).

Table 10 compares the ability to preserve cranial nerves for each tumor class. Overall, the hypoglossal nerve was preserved in 78 (74%) patients, the accessory

Table 5. Distribution of Temporal Bone Paragangliomas by Class

Class	*Number of Patients*	*Percentage*
A	8	6
B	5	4
C	33	25
C+D	86	65
Total	132	100

Table 7. Distribution of Cranial Nerve Deficits by Tumor Class (n, %)*

Tumor Class	*N*	*No Deficits*	*VI*	*VII*	*VIII†*	*IX*	*X*	*XI*	*XII*
A	8	8 (100)	—	—	—	—	—	—	—
B	5	5 (100)	—	—	—	—	—	—	—
C1	14	5 (42)	—	2 (17)	4 (31)	2 (18)	2 (18)	3 (25)	1 (9)
C2	40	8 (20)	—	13 (33)	17 (45)	7 (19)	9 (24)	4 (11)	8 (21)
C3	45	5 (11)	2 (4)	20 (44)	25 (56)	20 (44)	26 (58)	15 (33)	22 (49)
C4	20	1 (5)	4 (20)	17 (85)	17 (85)	16 (80)	16 (80)	14 (70)	10 (50)
Total	132	32 (24)	6 (5)	52 (39)	63 (48)	45 (34)	53 (40)	36 (27)	41 (31)

*not all patients available for evaluation
†cochlear division

Table 8. Surgical Approaches Undertaken by Tumor Class (n = 132; n, %)

Class	*N*	*Transcanal*	*Transmastoid*	*SP**	*Type A†*	*Type A + B‡*
A	8	3 (37)	5 (63)	—	—	—
B	5	—	2 (40)	2 (40)	1 (20)	—
C1	14	—	2 (14)	2 (14)	10 (72)	—
C2	40	—	—	1 (2)	39 (98)	—
C3	45	—	—	—	45 (100)	—
C4	20	—	—	—	16 (80)	4 (20)
Total	132	3 (2)	9 (7)	5 (4)	111 (84)	4 (3)

*Subtotal petrosectomy
†Infratemporal fossa approach type A
‡Combined infratemporal fossa approaches types A & B

Table 9. Operation Time, Estimated Blood Loss, and Hospitalization (n = 132; Average, Range)

Class	*N*	*Operation Time (h)*	*Estimated Blood Loss (L)*	*Hospitalization (d)*
A	8	1.4 (0.25–3.0)	0.2 (0–1.0)	8 (5–12)
B	5	3.5 (2.0–5.6)	0.4 (0–1.0)	13 (5–12)
C1	14	5.4 (2.5–9.0)	1.9 (0–8.0)	16 (6–38)
C2	40	6.3 (4.0–9.3)	1.6 (0.3–4.0)	20 (9–65)
C3	45	6.5 (2.6–9.0)	2.4 (0.7–11)	20 (9–44)
C4	20	7.2 (5.2–8.5)	4.6 (1.2–10)	20 (11–29)
Total	132	6.1 (0.25–9.3)	2.0 (0–11)	18 (5–65)

Table 10. Preservation of Lower Cranial Nerves (%)

Class	*VIII (n = 84)*	*IX (n = 85)*	*X (n = 118)*	*XI (n = 111)*	*XII (n = 106)*
A	100	100	100	100	100
B	100	100	100	100	100
C	56	44	79	94	83
CDe	31	21	63	86	81
CDeDi	27	2	21	4	55
Total	48	41	54	71	74

Table 11. Facial Nerve Management

Method	Number of Patients	%
Intact fallopian canal	16	13
Permanent anterior displacement	91	72
Intact epineurium	65	
Segmental resection of epineurium	26	
Cable graft	10	8
VII–XII anastomosis	6	5
VII–XI anastomosis	1	1
VII–V anastomosis	1	1
No reconstruction	1	1
Total	126	100

nerve in 79 (71%) patients, the vagus nerve in 64 (54%) patients, the vestibulocochlear in 40 (48%) patients, and the glossopharyngeal nerve in 33 (41%) patients. The ability to preserve the nerve was directly proportional to the size of tumor, with lesser outcomes when intradural tumor was encountered. The vestibulocochlear nerve was considered to be preserved if the otic capsule and the internal auditory canal were not destroyed during surgery.

The surgical management of the facial nerve is outlined in Table 11. Facial nerve integrity was preserved in 107 (89%) of 126 patients with complete data, whereas others underwent either cable grafting (n = 10, 8%), XII to VII anastamosis (n = 6, 5%), XI to VII anastomosis (n = 1, 1%), or V to VII grafting and anastomosis (n = 1, 1%). One patient who had had three previous attempts at tumor resection and had lived with a complete facial paralysis for 10 years refused any attempt at facial nerve reconstruction after tumor excision. Sixty-nine patients were followed for facial nerve function for an average of 3.4 years (range 2 to 11) following surgery. Excellent facial nerve function (Fisch grade 76 to 100%, House-Brackman grade I to II) was obtained in 81% of these patients overall. The best possible outcome was of course obtained in those patients in whom the nerve was not disturbed, with 100% normal function postoperatively. Of the 52 patients who underwent anterior displacement, 46 (88%) had Fisch grade 76 to 100%/House-Brackman grade I to II function. Of those patients who underwent grafting or crossover, 7 of 8 (88%) had Fisch grade 51 to 75%/House-Brackman grade III, and 1 of 8 (12%) had Fisch grade 26 to 50%/House-Brackman grade IV. No patients had an outcome of less than Fisch grade 26 to 50%/House-Brackman grade IV. The conversion of Fisch grade to House-Brackman grade was made according to a table published by House in 1985. According to this conversion table, patients with good eye closure and sphincter control are classified House-Brackman III (Fisch grade 51 to 75%) in spite of the limited forehead motion. The data are summarized in Table 12.

Total tumor removal was achieved in 109 patients (83%) (Table 13). This included patients with tumor extension significantly within the dura. Of those patients with Di1 tumors (up to 2-cm diameter intradural extension), 28 of 35 (80%) were totally excised; 8 of 23 (35%) Di2 tumors with a 2-cm or more intradural extent could be totally removed. In 22 cases (17%) tumor extirpation was limited by tumor involvement significantly beyond the dura. In only 1 case was subtotal tumor removal performed because of intracranial tumor that did not penetrate the dura (class C3De2) because of extensive involvement of both ICAs. No patient underwent limited removal (enlarged biopsy). The degree of tumor removal was confirmed 2 to 12 weeks postoperatively by neuroradiological evaluation. In no case was residual tumor discovered by postoperative neuroradiology that was not known at the time of surgery.

Complications

Postoperative complications are listed in Table 14. Cerebrospinal fluid (CSF) leak was the most common (11%), and its occurrence was most frequent in the class D tumors (Table 15). A frequent source of CSF leak was the defects made in the posterior fossa dura when passing the suture around the sigmoid sinus during its liga-

Table 12. Postoperative Facial Nerve Function (Fisch Grade / House-Brackman Grade) (in Percent)

Fisch House Procedure	N	100% I	99–76% II	75–51% III	50–26% IV	25–1% V	0% VI
Left In Situ	9	100	—	—	—	—	—
Ant Displ*	43	68	19	11	2	—	—
Ant Displ SR†	9	78	22	—	—	—	—
Cable graft	5	—	—	80	20	—	—
VII–XII	2	—	—	100	—	—	—
VII–V	1	—	—	100	—	—	—
Total	69	67	14	16	3	—	—

*Displ, Displacement
†SR, Segmental resection of epineurium

Table 13. Influence of Tumor Class on Extent of Removal

Class	*Number of Patients*	*Total Excision N (%)*	*Subtotal Excison N (%)*	*Partial Excision N (%)*
A	8	8 (100)	—	—
B	5	5 (100)	—	—
C1	11	11 (100)	—	—
C1De1	3	3 (100)	—	—
C2	16	16 (100)	—	—
C2De1	15	15 (100)	—	—
C2De1Di1	7	6 (86)	1 (14)	—
C2De2Di1	2	1 (50)	1 (50)	—
C3	5	5 (100)	—	—
C3De1	7	7 (100)	—	—
C3De1Di1	7	6 (86)	1 (14)	—
C3De1Di2	2	0	2 (100)	—
C3De2	2	1 (50)	1 (50)	—
C3De2Di1	15	13 (87)	2 (13)	—
C3De2Di2	7	3 (43)	4 (57)	—
C4	1	1 (100)	—	—
C4De1	1	1 (100)	—	—
C4De1Di1	1	1 (100)	—	—
C4De2Di1	3	1 (33)	2 (67)	—
C4De2Di2	14	5 (36)	8 (57)	1 (7)
Total	132	109 (82)	22 (17)	1 (1)

tion. Since we began incorporating a small muscle graft in the ligature to seal the defect, we have had no further problems. Almost as common was postoperative aspiration, noted in 10% of cases, although in no instance was tracheotomy required. Other than 8 cases of local wound infection (6%), the other complications (pulmonary embolus, meningitis, and stroke) were isolated occurrences. There was only 1 death in this series, the result of a cardioplumonary arrest on postoperative day 10.

Follow-up

Eighty-three patients were followed for the status of tumor removal for an average of 2.1 (range 2 to 11) years after surgery (a significant number of our patients come from other countries and can be difficult to follow long term). Of 63 patients who had total tumor removal, 62 (98%) were found to be tumor free on subsequent clinical and neuroradiological evaluation. One patient who initially had a class C tumor had a recurrence 10 years after surgical removal. All 19 of those having had subtotal removal were found to have residual tumor consistent with that found on their initial postoperative neuroradiological examinations. None of them showed evidence of aggressive tumor behavior on these investigations. In the 1 patient who had had a partial tumor removal, there was no deterioration of clinical or neuroradiological status from the first postoperative evaluation at the time of his last follow-up.

DISCUSSION

The overall patient population in this study was typical for paraganglioma. The ratio of female to male patient was practically 2:1, without a predominance for the right or left side.[9,10] The 5% incidence of a familial inheritance was somewhat less than the usual 10%,[11] that is thought to be transmitted through chromosome 11q.[12] These tumors, as expected, tended more often to be multicentric (43%) than were those that were sporadic (14%).[12,13] Unusual in this study was that 71% of the inherited tumors were in men, rather than the equal distribution previously reported.[15] The inherited tumors

Table 14. Postoperative Complications (n = 132)

Complication	*N*	*%*
CSF Leak	14	11
Aspiration	13	10
Wound infection	8	6
Pulmonary embolus	2	2
Meningitis	1	1
Cerebrovascular accident (stroke)	1	1
Cardiopulmonary failure (death)	1	1

Table 15. Tumor Class and CSF Leak

Class	*Number of Patients*	*CSF Leak*	*%*
A	8	0	0
B	5	0	0
C	33	3	8
CDe	28	5	20
CDi	58	6	11
Total	132	14	11

also tended to occur in younger patients (mean of 32 years versus 44 years in sporadic tumors). Our data would suggest the screening of the offspring, especially male, of those with familial tumors. The ideal time for this would be in young adulthood.

This series is also typical in the 2- to 3-year delay between the onset of symptoms, referral, and diagnosis.[16,17] This is due to both the slow growth of these tumors and the vague symptoms, for example, tinnitis, with which they often present.

Among the unique aspects of this series are the number of patients (the largest in the literature) and the highly advanced class of their tumors (90% C or C+D, 65% with intracranial extension). The fact that our treatment protocol has changed little over the past 25 years has allowed us to critically analyze the outcome of our management with extensive data. The key aspects of the infratemporal fossa approaches are that through permanent anterior transposition of the facial nerve, direct access is provided to the most extensive tumors deep within the temporal bone from the jugular bulb and jugular foramen to the pyramid apex. Safety is afforded in the capacity to control and manipulate the internal carotid artery. This is reflected in our ability to fully resect (confirmed by MRI or CT) these tumors in 83% of the cases, with a postoperative complication rate of 11%, a mortality rate of 0.7%, and a 98% tumor-free survival in long-term follow-up. Furthermore, in 80% of patients with intradural extension of up to 2-cm complete single-stage tumor excision was possible.

It is interesting that in those patients with less than total tumor excision (but less than 10% of tumor volume remaining), long-term clinical and neuroradiological follow-up showed no progression of disease. Through this subtotal surgery, these tumors are essentially devascularized and thus deprived of the ability to grow. When tumor is left intradurally, the core of the intracranial residual is cauterized, thus destroying the blood supply.

The topic of cranial nerve preservation is central in the discussion of therapy of paragangliomas of the temporal bone. As a quarternary referral center, the patients referred to us tend to have far advanced tumors. Only 10% of cases were class A or B; all of these were fully excised, and none had postoperative cranial nerve deficits. Of the remaining patients who had class C or C+D tumors, 84% (100 of 119) had at least one preoperative cranial nerve deficit. Furthermore, neural invasion by tumor was found to be more extensive than would be predicted by normal preoperative neural function. (Fifty percent of nerves showing evidence of invasion intraoperatively have been previously shown to have normal function preoperatively.[18]) The rate of lower cranial nerve preservation in this study varied between 31 and 70%. In those patients for whom all cranial nerve function could not be spared, the morbidity was often quite minimal. When cranial nerve dysfunction develops slowly, the patient adapts well. When the nerve must be sacrificied intraoperatively, the compensation can be more difficult. Perhaps the most difficult situation is when functional nineth and tenth cranial nerves must be resected in an elderly patient. This is, however, rarely the case because nerve function has already been compromised by the tumor in most of these patients. Even in extreme cases tracheotomy was avoided by the use of parenteral nutrition, and swallowing was begun on the fourth to sixth postoperative day. Through the use of postoperative swallowing therapy, oral feeding was resumed within the first week after surgery in all cases.

The topic of facial nerve preservation deserves special mention. To gain full access to the jugular bulb, jugular foramen, intratemporal carotid artery, and infiltrated temporal bone, it is necessary to displace the facial nerve anteriorly. To minimize damage to the nerve in this process, we use constant nerve monitoring. The nerve is separated sharply from its medial vascular and fibrous attachents. The nerve itself is not grasped, rather it is manipulated by its encompassing connective tissue, with cottonoids and a special microraspatory. At the stylomastoid foramen, where the nerve is tightly adherent to the surrounding dense connective tissue, it is mobilized en bloc without exposing its epineurium. The nerve is then carefully mobilized and transposed into its neocanal and groove in the upper parotid, where it is sutured gently in place in a cradle of parotid tissue. The nerve must never be stretched during the procedure, and the nerve monitor should register contact potentials but never irritation potentials.

The most common area of the nerve to be involved in tumor is the mastoid segment, at its medial surface. A grading scheme for neural invasion has been published previously.[18] With grade I tumor involvement (tumor remains 1 mm or more from the perineurium), the nerve can be separated from the tumor bulk with the epineurium intact. When the epineurium is invaded (grade II), it can be resected, leaving the perineurium undisturbed. This must be done after anterior transposition and tumor removal to avoid injury of the weakened nerve within its parotid cradle. When the perineurium (grade III) or the endoneurium (grade IV) is invaded, the segment of nerve must be resected. Reconstruction is performed by grafting when possible; if the proximal nerve stump is not suitable because of location or poor condition, a hypoglossal-facial crossover is performed. When the hypoglossal nerve is not usable, we then perform either an accessory-facial crossover or trigeminal-facial crossover graft.

Unfortunately, without the benefit of a randomized prospective trial it is not possible to lay to rest the controversy of surgery versus radiation as the optimal primary therapy for paragangliomas of the temporal bone; however, we can present the logic behind our treatment protocol. Paragangliomas are benign, slow-growing tumors; radiation is most effective for rapidly growing tumors with a high mitotic index. Radiation does not seem

to affect the neoplastic component of the tumor (chief cells) that persists chronically after therapy and may continue to secrete catecholamines.[19–21] Radiation causes diminution of the small vessels, decreased arteriovenous shunting, and proliferative endarteritis,[21–23] but does not seem to affect the larger vessels[24] and causes no overall decrease in tumor vascularity. The overall effect seems to be proliferation of the fibrous stroma.[23,25] The tumor mass rarely decreases after radiation and may in fact continue to increase.[26–29] Radiation does not prevent infiltration of neural structures.[18] Furthermore, the complications of radiation therapy may include temporary hair loss, mucositis, serous otitis media, stenosis of the external auditory canal, chronic otitis media, osteoradionecrosis, neuropathy of the fifth through twelfth cranial nerves, brain necrosis, and radiation-induced neoplasm or malignant transformation that arises on average 8 to 10 years after therapy.[1,10,30–36] We would agree with Carrasco and Rosenman ,[37] that "control of tumor by surgical eradication rather than alteration of biologic potential with radiation seems safer, especially in the younger patient." Because the average age of the patients in our study was 44 years, this is a significant consideration. Thus, it is our policy that radiation is not recommended as primary therapy unless surgery is contraindicated for medical reasons. This is especially true for class A and B tumors, for which the morbidity and mortality of surgery is close to zero and the recurrence rate is negligible.

In reviewing the changes in our management of temporal bone paragangliomas since the 1982 series report, several points should be mentioned. We have added to the classification scheme the class C4, representing tumor involvement of the foramen lacerum and cavernous sinus, to aid in the discussion of the most advanced tumors. We have added differentiation in the intracranial class D tumors into De (intracranial, extradural tumor) and Di (intracranial, intradural extension), with subclasses 1 (up to 2-cm diameter) and 2 (more than 2-cm diameter), for more precision in documentation and analysis. Although embolization was previously felt to be too risky in relation to its benefit in diminishing intraoperative hemorrhage, its safety has been improved to the point that we now use it routinely. In addition, when preoperative analysis suggests extensive involvement of the ICA with tumor in a young patient and resection of the vessel may be necessary for erradication of disease, a preoperative balloon occlusion test may be performed so that intraoperative balloon occlusion is an option when deemed necessary. To avoid postoperative CSF leak when repairing dural defects such as those that occur when ligating the sigmoid sinus, we also include a graft of muscle in the closure to ensure a tight seal. The edges of the dura are sutured to the skull to avoid the formation of an epidural hematoma.

To conclude, the results of this series of 136 patients treated by the same surgical protocol demonstrate the safety and efficacy of surgical treatment of even highly advanced paragangliomas of the temporal bone, with a disease-free long-term survival rate of 98% in those patients who had total resection. With the infratemporal fossa approaches, our ability to resect a tumor is limited no longer by tumor size but by the magnitude of intradural extension. Even in those patients in whom intradural tumor is not fully resected (class Di2), cauterization of the dural blood supply has prevented further tumor growth. The ability to preserve the lower cranial nerves depends on tumor size, but the morbidity from their resection, when necessary, is not prohibitive. We have found that even though subtotal petrosectomy causes a conductive hearing loss, this procedure is necessary for patient safety in tumors of class C1 or larger. Postoperative CSF leakage and subsequent meningitis are prevented, and the necessary access for control of tumor invasion is obtained. Thus, with minimal morbidity and a mortality rate of less than 1%, we have confirmed that surgical therapy is the first line treatment for paragangliomas of the temporal bone.

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