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Management of Complex Tympanojugular Paragangliomas Including Endovascular Intervention

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Objectives/Hypothesis: To identify complex cases of tympanojugular paraganglioma (TJP) and to analyze their management and surgical outcome. To present strategy and guidelines on how to deal with the perioperative complicating factors.

Study Design: Retrospective analysis.

Methods: We retrospectively studied 212 TJP patients to identify the complex cases and analyzed their management and surgical outcome. The criteria for complex TJP are: 1) very large size; 2) large intradural extension (IDE); 3) extension to the cavernous sinus, foramen magnum, and clivus; 4) significant involvement of the internal carotid artery (ICA); 5) a solitary ICA on the lesion side; 6) involvement of the vertebral artery; 7) dominant or unilateral sigmoid sinus on the lesion side; 8) bilateral or multiple paragangliomas; and 9) recurrence.

Results: Sixty patients were classified as complex. Forty-five patients had tumors with IDE. Twenty patients with tumor involving ICA underwent preoperative endovascular intervention. Two patients had a solitary ICA and two others unilateral sigmoid sinus on the lesion side. Thirteen patients had multicentric tumors, and three patients had bilateral ones. The main procedure was the infratemporal fossa approach type A. Total removal was achieved in 46 patients, and five of them experienced recurrence.

Conclusions: It is essential to carefully investigate the hemodynamics of the brain in planning surgery. A proper preoperative endovascular intervention facilitates gross total tumor removal. In bilateral paragangliomas, lower cranial nerves' function should be preserved at least unilaterally. Staged removal is recommended for a tumor with a large intradural component.

Key Words: Paraganglioma, glomus jugulare tumor, chemodectoma, internal carotid artery, vertebral artery, skull base neoplasms, facial nerve.

Level of Evidence: 4.

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INTRODUCTION

Tympanojugular paragangliomas (TJP) are histologically benign and slow growing tumors, but they can be locally aggressive and infiltrate the skull base.[1] Total surgical extirpation is the mainstay of treatment for TJP.[2] Concurrent advances in techniques of imaging, intervention, anesthesia, and intensive care have made surgery safer and more effective.[3–6]

However, some TJP cases are still challenging to treat.[3,7–12] Examples of such cases include large tumors, tumors with large intradural extension, tumors involving the cavernous sinus, internal carotid artery (ICA) or vertebral artery (VA), and previously operated or irradiated tumors. Also included are tumors with a single carotid artery, dominant or unilateral sigmoid sinus on the side of the lesion, or bilateral or multiple tumors. It is difficult to decide the treatment for these tumors, and accurate preoperative surgical planning is mandatory. We usually carry out the standard preoperative evaluation, which includes temporal bone computed tomography (CT), gadolinium-enhanced magnetic resonance imaging (MRI), and angiography, with the study of the arterial and venous phase. Though the management of the ICA/VA in complex cases has not been established yet, stent insertion or permanent balloon occlusion (PBO) is required in certain cases.[13] Staged removal is performed for cases with a large intradural extension.[1,3] For other complex cases, in which the tumor reaches particular areas such as the cavernous sinus or the foramen magnum, radiotherapy may be necessary after preplanned subtotal removal.

In this article we present the management and surgical outcomes in complex TJP, and suggest a new stage for the Fisch classification[3,14] for the VA involvement.

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MATERIALS AND METHODS

We present a retrospective study of 60 patients with complex TJP out of 212 patients who were treated at the Gruppo Otologico in Piacenza, Italy between 1988 and 2009. The

TABLE I.
Distribution of Patient With Complex Tympanojugular Paragangliomas.

Factors of Complex TJP	No. of Cases
Very large tumor	
C3*	27
C4*	5
De2*	3
Intradural extension	
Di1*	25
Di2*	18
Di3*	2
Extension to	
Cavernous sinus	2
Foramen magnum	3
Clivus	8
Serious involvement of	
ICA	20
VA	4
A single ICA on the lesion side	2
Unilateral sigmoid sinus on the lesion side	2
Dominant sigmoid sinus on the lesion side	1
Malignancy (or very aggressive case)	1 (or 1)†
Bilateral paragangliomas	3
Multiple paragangliomas	11
History of previous treatment	13
Total	60

*According to the Fisch classification.[15]

†Very aggressive case-like malignant biologic behavior in spite of histologically benign paraganglioma.

TJP = tympanojugular paraganglioma, ICA = internal carotid artery, VA = vertebral artery.

criteria for complex TJP are as follows: 1) a very large size (Fisch classification C3-4, De2, with Di1-3)[15]; 2) a large intradural extension; 3) involvement of the cavernous sinus, foramen magnum, clivus, or VA; 4) multiple or bilateral paragangliomas; 5) with unilateral or dominant sigmoid sinus on side of the lesion; 6) with a single ICA on the side of the lesion; and 7) recurrence after previous surgery or radiotherapy. All patients underwent pure-tone audiometry, lower cranial nerve (LCN) function examination with fiberoptic laryngoscope, imaging studies including temporal bone CT, gadolinium-enhanced MRI and four-vessel angiography with manual cross-compression testing, and superselective tumor embolization 2 days before surgery in every case.[16,17]

Preoperative endovascular intervention of the ICA included PBO and stenting. Indications for preoperative endovascular intervention of the ICA were according to MRI and angiographic data and included[13]:

1. Encasement of the distal cervical, vertical, and horizontal segments of the ICA between 270° and 360°, as shown by CT and MRI in the axial plane.
2. Evidence of stenosis and irregularities of the arterial lumen as determined by angiography.
3. Extensive blood supply from ICA branches as seen on angiography.
4. Previous surgery with ICA manipulation and/or previous radiotherapy.

We performed a manual cross-compression internal carotid artery occlusion test in every case. In cases where carotid stenting was not possible and there was sufficient collateral cerebral circulation confirmed by manual cross-compression testing, permanent balloon occlusion was performed. In cases where carotid stenting was not possible and there was insufficient collateral cerebral circulation, the lesion around the internal carotid artery was conservatively removed.

The facial nerve function was recorded using the modified House-Brackmann (HB) grading system.[18] The main procedure was performed using the infratemporal fossa approach type A (IFTA-A).[15,17] Supplementary techniques were used for some patients. The patients were followed for a minimum of 1 year. Subsequently, the patients underwent an annual radiological follow-up (CT scan and MRI) to detect any recurrence for at least 10 years.

RESULTS

There were 39 females and 21 males. Their ages ranged from 16 to 73 years with a mean age of 42.4 years. Thirty-two patients had tumors on the left side, and 28 on the right. Forty-five patients had tumors with an intradural extension. The cavernous sinus was involved in two patients, the foramen magnum in three patients, and the clivus in eight patients. Among 20 patients with ICA (C3-C4) involvement, nine underwent PBO and 11 intraluminal stent insertion. Four cases had tumors involving the VA, and two of them received permanent occlusion. Two patients had a single ICA on the side of the lesion, and three patients had an unilateral or dominant sigmoid sinus on the lesion side of the lesion.

Eleven patients had multicentric paragangliomas and three bilateral paragangliomas. Table I summarizes patient distribution of complex TJP. Thirteen patients had been previously treated as shown in Table II. The presenting symptoms and signs are listed in Table III. The most common cranial nerve deficit (CND) was found on IX and X nerves (Table IV). LCN functional preservation rate was 74.1% (Table V).

In 45 patients with no facial palsy, anterior rerouting of the facial nerve was performed, and its results are summarized (Table VI). Of these, 29 patients (66.7%) had HB grade I or II at 1 year follow-up after surgery. In four patients with incomplete facial nerve palsy, anterior rerouting of the facial nerve was done. In nine patients, the facial nerve was sacrificed due to tumor-infiltration, and immediate nerve grafting was performed. In two of them, nerve grafting was not performed due to the failure of the identification of

TABLE II.
Types of Previous Treatments.

Previous Treatment	No. of Cases
Mastoidectomy without anterior rerouting of FN	8
Infratemporal fossa approach type A	2
Fallopian bridge technique	1
Radiotherapy + mastoidectomy	1
Radiotherapy	1
Total	13

FN = facial nerve.

TABLE III.
Presenting Symptoms/Signs in Complex Tympanojugular Paragangliomas (N = 60).

	No. of Cases (%)
Hearing loss	48 (80.0)
Tinnitus	34 (56.7)
Hoarseness	13 (21.7)
Pulsatile tinnitus	13 (21.7)
Instability	8 (13.3)
Dizziness	6 (10.0)
Facial weakness	5 (8.3)
Sternocleidomastoid weakness	4 (6.7)
Dysphagia	3 (5.0)
Otorrhea	3 (5.0)
Tongue deviation	3 (5.0)
Diplopia	3 (5.0)
Decreased gag reflex	2 (3.3)
Otalgia	1 (1.7)
Headache	1 (1.7)

proximal facial nerve or the patient's refusal to undergo the second stage of surgery. The other two patients had no treatment for the facial nerve due to their complete facial nerve palsy (Table VII).

In 46 patients (76.7%), gross total removal was achieved. Recurrence occurred in five patients (11.1%). Among them, one with malignant TJP showed nodal recurrence 3 years after surgery, and revision surgery is planned. One patient presented with very aggressive tumor growth just after the second stage of surgery. Another patient had been operated on in 1975 at another institution, had recurrence and revision surgery in 1987 at our institution, and had another recurrence in 1998. The patient has been followed with serial imaging studies, and the recurrent tumor has not shown any growth yet. Two other patients had recurrences 8 and 13 years after the initial procedure, but the recurrence has been stable so far. In cases with planned staged operations at our institution we experienced only negligible bleeding. However, in recurrent cases after incomplete surgery by another institution, we occasionally experienced moderate bleeding of less than 250 mL during dissection due to anatomical distortion by previous surgery. None of our cases required a blood transfusion. Table VIII lists the causes for subtotal resection.

Postoperative complications are summarized in Table IX.

TABLE IV.
Outcomes of Pre- and Postoperative Cranial Nerve Function in 60 Cases.

	No. of Cases With Cranial Nerve Deficit (%)	
Cranial Nerve	Preoperative	Postoperative*
IV	2 (3.3)	1 (1.7)
V	1 (1.7)	—
VI	3 (5.0)	2 (3.3%)
IX	26 (43.3)	41 (68.3)
X	25 (41.7)	35 (58.3)
XI	15 (25.0)	25 (41.7)
XII	16 (26.7)	22 (36.7)

*At least 1 year after the initial procedure.

TABLE V.
Preservation Rate of Lower Cranial Nerve Function in 60 Cases.

Cranial Nerve	Nerves Preserved*/ Clinically Uninvolved Nerves†	%
IX	19/34	55.9
X	25/35	71.4
XI	35/45	77.8
XII	38/44	86.4
Total	117/158	74.1

*At least 1 year after surgery.
†Preoperative status.

Clinical Cases With Illustrations

Case 1. A 57-year-old female presented to our institution with a 3-year history of unilateral hearing loss, dysphonia, and aspiration of liquid. Otoscopy revealed a polyp-like mass occupying the medial compartment of the external auditory canal. Examination of cranial nerves (CNs) revealed left X and XII CN palsy. On audiogram, she had a left dead ear. The MRI showed a left Fisch class C3Di2 TJP[15] and Fisch stage II_m vagal paraganglioma[19] (Fig. 1A, 1B, 1C). Angiography showed severe stenosis and anterior displacement of the cervical ICA (Fig. 1D). To determine the efficacy of collateral circulation, preoperative occlusion test was done with manual cross compression of the ipsilateral ICA. Due to severe stenosis of the ICA, PBO was decided on instead of stent insertion. To permit the adaptation of cerebral vasculature to the new hemodynamic situation, the surgery was postponed for 1 month. After superselective embolization, IFTA-A was performed. The bony labyrinth was left for the landmark of the planned second-stage surgery. Tumor-infiltrated LCNs were sacrificed. Due to the tumor-infiltrated periosteal sheath of the ICA, extensive bone removal was performed from the horizontal segment to the foramen lacerum. The cervical ICA was narrowed and displaced by a large vagal paraganglioma.

The ICA was doubly closed with vascular clips just above the carotid bifurcation and transected at the level of the foramen lacerum and just above the carotid

TABLE VI.
Functional Outcomes of Facial Nerve After Anterior Rerouting in 45 Cases With Preoperatively Intact Facial Nerve Function.

	No. of Cases		
House-Brackmann Grade*	Preoperative	Postoperative (%)	
I	45	12 (26.7)	66.7 (I + II)
II	—	17 (40.0)	
III	—	14 (33.3)	

*According to the House-Brackmann grading system.[18]

TABLE VII.
Surgical Outcomes of Facial Nerve Function in 15 Cases That Underwent Nerve Graft, Sacrifice, or Had Abnormal Preoperative Facial Function.

Case	Procedures	Treatments of FN	Pre-op FN Function*	Post-op FN Function*
1	IFTA-A	Nerve graft	I	III
2	IFTA-A	Nerve graft	VI	IV
3	IFTA-A	Nerve graft	I	IV
4	IFTA-A	Nerve graft	II	V
5	IFTA-A+TLA	Nerve graft	III	III
6	TC	Nerve graft	I	IV
7	IFTA-A	Nerve graft	I	III
8	IFTA-A+TC	Sacrifice	I	VI
9	IFTA-A+EL	Sacrifice	IV	VI
10	IFTA-A	AR of FN	II	I
11	IFTA-A+TLA	AR of FN	IV	IV
12	IFTA-A+TLA	AR of FN	IV	III
13	IFTA-A+TLA	AR of FN	IV	IV
14	IFTA-A+TC	No treatment	VI	VI
15	TC	No treatment	VI	VI

*According to the House-Brackmann grading system.[18]
FN = facial nerve, IFTA-A = infratemporal fossa approach type A, TLA = translabyrinthine approach, TC = modified transcochlear approach, EL = extreme lateral transcondylar approach, AR = anterior rerouting.

bifurcation. The extradural part of TJP and vagal paraganglioma were simultaneously removed. MRI after first-stage surgery showed a residual intradural component of the tumor (Fig. 1E). In the second-stage surgery, the petro-occipital trans-sigmoid (POTS) approach[17,20] and labyrinthectomy were performed, and the intradural portion of the tumor was completely removed. The postoperative MRI after the second-stage surgery revealed no residual tumor (Fig. 1F). The facial nerve function has recovered to HB grade III and swallowing function was well compensated. There is no evidence of recurrence after 4 years thus far.

Case 2. A 41-year-old male presented with symptoms of facial palsy and hearing loss. He had undergone the so-called functional operations three times at another institution. On physical examination, he had HB grade IV facial palsy, hypoglossal nerve palsy, and a dead ear. MRI showed a huge intradural extension of the tumor and severe involvement of the sigmoid and transverse sinuses, ICA, and VA (Fig. 2A, 2B). After PBO of the ICA, IFTA-A was performed. During the first-stage surgery, the extradural tumor was removed. The ICA was resected at the level of genu between the vertical and horizontal segments and just above the bifurcation. The infiltrated hypoglossal nerve was sacrificed, and both X and XI CNs were preserved. The tumor-infiltrated facial nerve at the stylomastoid foramen was sacrificed and could not be reconstructed because the proximal stump could not be identified. Before the second-stage surgery, the tumor-involved VA was permanently occluded with coils. Excision via a modified transcochlear approach (MTCA) was performed.[21,22] The clivus and atlanto-occiptal joint were partially drilled out. En bloc removal was achieved. MRI after the second surgery showed that an about 25 × 10 × 12 mm-sized tumor still remained at the foramen magnum. To remove this lesion, an extreme lateral transcondylar approach was performed. At the level of the second cervical vertebra, the VA was identified and followed. A tumor was identified near the foramen

TABLE VIII.
Analysis of Subtotal Resection Cases (n = 14 Cases).

Causes	No. of Cases	Follow-Up Period, Tumor Size, Progress in Each Case
No 2nd-stage operation or follow-up	6	1 year, follow-up loss; 5 years, 1 cm, stable; 1.5 year, 1.5 cm, stable; 1 year, follow-up loss; 1 year, 2 cm, no symptom; 1.5 year, follow-up loss
Extension to cavernous sinus	2	γ-knife, 6 years, 0.3 cm, stable; γ-knife, 9 years, tumor disappeared
Excessive bleeding at the clivus	2	3 years, 1 cm, stable; 10 years, 2.2 cm, no symptom
Excessive bleeding at the petrous apex	1	1 year, refusal of additional treatment
Excessive bleeding at the FM	1	2 year, 0.5 cm at the FM, stable
ICA treatment without pre-op	1	1 year, follow-up loss
VA treatment without pre-op	1	γ-knife, 3 years, stable
Total	14	

γ-knife = γ-knife surgery; FM = foramen magnum, ICA = internal carotid artery, VA = vertebral artery, pre-op = preoperative intervention.

TABLE IX.
Postoperative Complications in 60 Cases.

Complication	No. of Cases
Cerebrospinal fluid leak	1
Aspiration	—
Pneumonia	1
Meningitis	—
Wound infection	—
Peritonitis	1
Death	—

magnum, and subtotal removal was accomplished due to bleeding, which obscured the surgical field. MRI 3 years after surgery showed a very small remnant at the foramen magnum (Fig. 2C, 2D, 2E). The remnant has been stable so far. If it shows growth, radiotherapy will be performed.

DISCUSSION

It is difficult to completely eliminate TJPs due to their location and locally infiltrative behavior.[11] The ideal treatment is total resection, and thanks to recent advances in microsurgical and preoperative interventional radiologic techniques, tumors that were once considered inoperable in the past can now be removed.[23] However, it is still difficult to remove tumors and avoid permanent morbidity for major vessels, the CNs, and brain stem at the same time.[3,7–12] These tumors have one or more complicating factors as follows:

1. Very large size
2. Large intradural extension
3. Extension to the cavernous sinus, foramen magnum, and clivus
4. Significant involvement and infiltration of the ICA
5. A single ICA on the lesion side
6. Involvement of the VA
7. Dominant or unilateral sigmoid sinus on the lesion side
8. Bilateral or multiple paragangliomas
9. Recurrence after previous surgery, radiotherapy, or stereotactic radiosurgery

Very Large Size

As TJPs grow, they extend either into the carotid canal and the petrous apex or into the intradural space through the medial wall of the jugular bulb so that they can involve the lower cranial nerves. Fisch class C3 and C4 tumors are generally considered as large tumors.[3,24] As a rule, IFTA-A can be used for C2 tumors, and IFTA-A or IFTA-A combined with IFTA type B can be used for C3/C4 tumors. If the tumor involves the clivus, occipital condyle, or foramen magnum, additional procedures such as MTCA or the extreme lateral transcondylar approach are necessary. Tumor size affects the preservation rate of LCNs.[25,26]

A Large Intradural Extension

The incidence of intradural extension for paragangliomas ranges from 14% to 72%.[11,14,27] As the number of Fisch class C tumors increases, the probability of intradural extension rises.[1,10] The larger the intradural extension, the more frequently they involve LCNs and compress the brainstem.

The degree of dural involvement can also be difficult to assess both preoperatively and intraoperatively. Small areas of an infiltrated dura can be easily resected and repaired. If an extensive area is of concern, planned second-stage resection can be considered. Any undetected involvement can lead to extensive intradural recurrence.

In our experience, the cleavage between tumor and brain stem has been easily established due to the devascularization of the tumor at the first-stage surgery and subsequent shrinkage of the intradural mass.

Several researchers suggested that simultaneous resection of both extradural and intradural lesions is feasible, regardless of the degree of extension.[4,6,10,28] Sacrifices of LCNs can cause severe aspiration with continuous cough, and the resulting increase in intracranial pressure can give rise to cerebrospinal fluid (CSF) leaks.[25] We prefer a staged operation for tumors with more than 2 cm intradural extension, which is similar to other studies, to prevent postoperative CSF leaks, and one-stage removal for tumors with 1 cm or smaller intradural extension.[3,17] Although the mean rate of postoperative CSF leak in single-stage surgery is 5.2% (ranged from 3.8% to 33.3%),[6,10,29,30] there was only one case of CSF leak in our study. At the second stage, the approach is determined by the location and size of the residual tumor and the patient's hearing function. Although the POTS approach[17,20] is preferred in most cases, a modified transcochlear approach or an extreme lateral transcondylar approach may also be used.

The lack of preoperative CNDs does not correlate with intraoperative neural involvement.[26] In fact, an intradural extension usually indicates infiltration of the LCN.[1] Some authors advocate that some tumors adherent to LCNs should be left intact for preservation of LCN function.[31] However, if total removal is technically feasible in young patients, we prefer to sacrifice tumor-infiltrated LCNs to prevent recurrence. The large majority of cases in which LCNs have been sacrificed compensated very well with or without intensive rehabilitation.

We experienced 45 cases with intradural extension: 25 class Di1, 18 class Di2, and two class Di3. In 14 cases, staged operation was done to prevent a postoperative CSF leak.

Extension to the Cavernous Sinus, Clivus, or Foramen Magnum

If TJP involves the cavernous sinus, it should be intentionally left intact to avoid compromising cranial nerves III, IV, and VI. Two cases involved the cavernous sinus, and they were intentionally left intact to maintain the ocular mobility. After surgery, stereotactic radiotherapy was carried out. In both cases, radiologic imaging

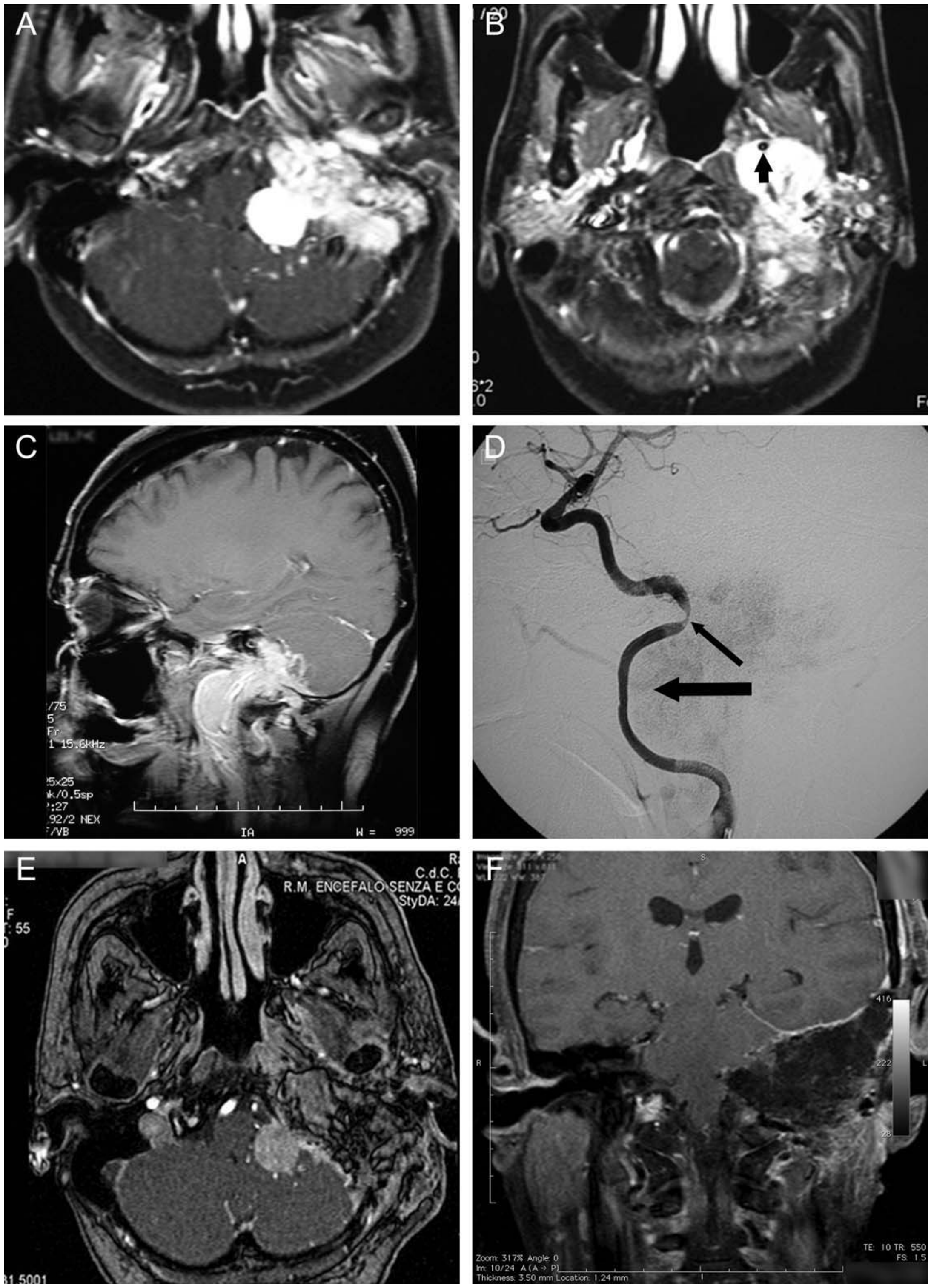

Fig. 1. Case 1 with the severe stenotic internal carotid artery (ICA) and multiple paragangliomas with a large intradural extension (Fisch class C3Di2). (A) Axial enhanced T1 magnetic resonance imaging (MRI) showing a large hypervascular mass at the level of the jugular bulb largely extending to the intradural space. (B) Enhanced T1 MRI revealing the severe stenosis of the cervical ICA (short arrow) compressed by a vagal paraganglioma. (C) Sagittal enhanced T1 MRI revealing a serious dural infiltration along the posterior fossa dura. (D) Digital subtraction angiography showing the cervical ICA that has been anteriorly displaced by the tumor (thick arrow) and is severely stenotic at the level of genu (thin arrow). (E) Enhanced T1 MRI after first-stage surgery. Extradural part of the tumor and the ICA were removed after permanent balloon occlusion. The intradural tumor is left for the staged operation. (F) Enhanced T1 fat suppression MRI after second-stage surgery. The dead space is filled with abdominal fat.

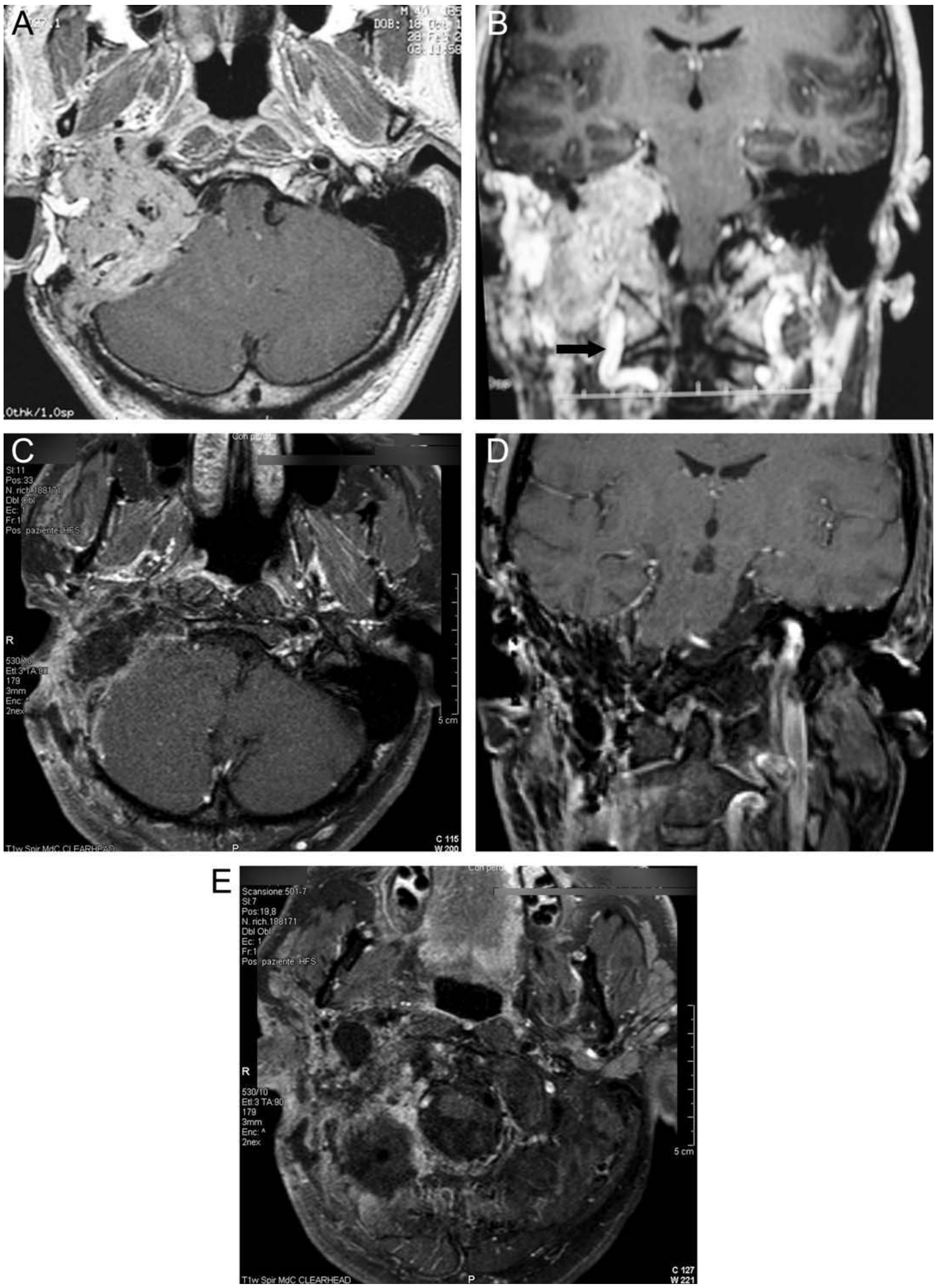

Fig. 2. Case 2 with a recurrent tumor (Fisch class C3Di2Ve) after three previous operations and involvement of the foramen magnum, clivus, internal carotid artery, and vertebral artery (VA). (A) Magnetic resonance imaging (MRI) demonstrating a huge tumor fully occupying the right infratemporal fossa, involving the internal carotid artery, and invading the clivus. (B) Coronal image revealing the right VA (black arrow) involvement. (C, D, E) MRI after third-stage surgery showing fat signal in the surgical defect. The small remnant tumor is noted around the foramen magnum.

showed no growth of the residual lesion in the cavernous sinus after 6 and 8 years postoperatively.

For tumors extending to the foramen magnum and lower clivus, MTCA type D or the extreme lateral approach may be used.[10,17] Any clival bone suspected of being infiltrated should be drilled out until healthy bone appears to prevent tumor recurrence.

We had three cases involving the foramen magnum, and total removal was difficult to achieve in one of them due to persistent bleeding.

Involvement of the Internal Carotid Artery

TJPs frequently involve the ICA due to their close anatomical proximity.[1] The degree of the involvement of the wall of the artery is an important factor that decides resectability of the tumor and affects surgical management. When indicated, the tumor must be dissected from the arterial wall. This could be done by subperiosteal (or supra-adventitial) or subadventitial dissection. Subperiosteal dissection is the separation of the layer between the adventitia and the periosteum of the carotid canal.[3] It is indicated when the tumor involves only the periosteum. This procedure is relatively easier and safer in the vertical segment than the horizontal segment, because the former is thicker and more accessible. Gentle displacement of the vertical segment of the artery is often required to remove the tumor that has extended anterior to the ICA. This requires mobilization of the artery from the entrance of the artery at the skull base to at least the genu of the horizontal segment. The caroticotympanic artery, arising just proximal to the internal carotid artery genu, is often responsible for a reasonable tumor blood supply, and care must be taken not to avulse this vessel during mobilization. Subadventitial dissection is the separation between the adventitia and the muscular layer. It can cause tears of the ICA wall, and therefore should be performed very carefully. The wall of the ICA at the level of the vertical segment is 1.5 to 2.0 mm thick with the adventitia being approximately 1 mm thick and is absent at the level of the horizontal portion.[24] Therefore, subadventitial dissection can be executed only at the level of the vertical portion.

Resection of the ICA may be required when it is completely surrounded by a tumor.

When the artery is completely covered by a tumor with severe stenosis on arteriography, manipulation without proper endovascular intervention may give rise to severe bleeding, incomplete removal, or cerebral vascular accident.[13,31–34] PBO is performed when the ICA is infiltrated by the tumor and the collateral blood flow is sufficient. In cases with insufficient collateral blood flow, we regularly use intraluminal stenting (Fig. 3A, 3B, 3C).[35] It reinforces the artery so that the surgeon can easily establish a cleavage plane on the external surface of the stent and more aggressively remove the tumor without the risk of blowout. The presence of the stent allows the safe mobilization of the artery if necessary. This new technique can allow reappraisal of selected cases previously suited only for subtotal resection.

A Single Internal Carotid Artery on the Lesion Side

As mentioned above, management of the ICA is essential for total tumor removal. For a case with a single carotid artery on the lesion side, possible options are "wait and scan," partial resection with radiotherapy, or total removal subsequent to the preoperative reinforcement with stents.[36] In a patient with a single carotid artery on the lesion side, bypass surgery can cause severe cerebral ischemic damage. Therefore, stent insertion may be the best option.

We have so far treated two patients with a single ICA without any problems. One of them was a 55-year-old female who had a previous contralateral ICA occlusion for the management of an intracranial aneurysm. Tumor infiltration of the vertical segment of the ICA was suspected on angiography. During the waiting and scanning period, tumor growth was noted. Although we discussed the option of preoperative high flow carotid artery bypass, our successful experiences with stent insertion led to us to proceed with stenting. Considering her age, the extent of the tumor, and brainstem compression radiotherapy was not indicated. After stent insertion, gross total tumor removal was performed. The patient had no cerebral vascular accident postoperatively.[36]

Involvement of the Vertebral Artery

To the best of our knowledge, excluding our cases, there were three cases of a tumor involving the VA reported in English journals.[10,37,38] When deciding the surgical method for TJPs, the VA must also be investigated to prevent a serious cerebellar vascular accident. We suggest adding another stage in the case of extradural and/or intradural vertebral artery involvement to the Fisch classification of TJP.[14]

We experienced four cases with a tumor involving the VA. In one case, we successfully removed the tumor from the VA without preoperative intervention. In another case, we failed to locate the surgical plane between the tumor and the artery. In the third case, we performed preoperative balloon occlusion and subsequently removed the extradural VA along with the tumor without vascular complication. In the fourth case, preoperative coil occlusion of the VA was carried out so that the tumor could be easily separated from the VA.

The preoperative evaluation and intervention of VA facilitates gross total tumor removal without serious surgical complications.

Dominant or Unilateral Sigmoid Sinus on the Lesion Side

Because TJP usually involves a jugular bulb, the closure of the sigmoid sinus and the ligation of the jugular vein are usually performed. However, closure of the dominant or unilateral sigmoid sinus on the lesion side may cause intracranial hypertension and venous congestion with brain swelling.[10] Therefore, preoperative evaluation of venous drainage of the brain is essential, including the ipsilateral mastoid emissary vein or condylar vein. If their diameters are bigger than normal, they should be preserved during surgery, closing the sigmoid sinus distal to the exit of the mastoid emissary vein. When collateral venous drainage cannot be preserved or when the patient has no sufficient collateral venous drainage, more conservative treatment such as partial resection with preservation of the sigmoid sinus, gamma knife surgery, or wait and scan is recommended.

Bilateral or Multiple Paragangliomas

Management of bilateral paragangliomas has a considerable and direct effect on the patient's life. Due to

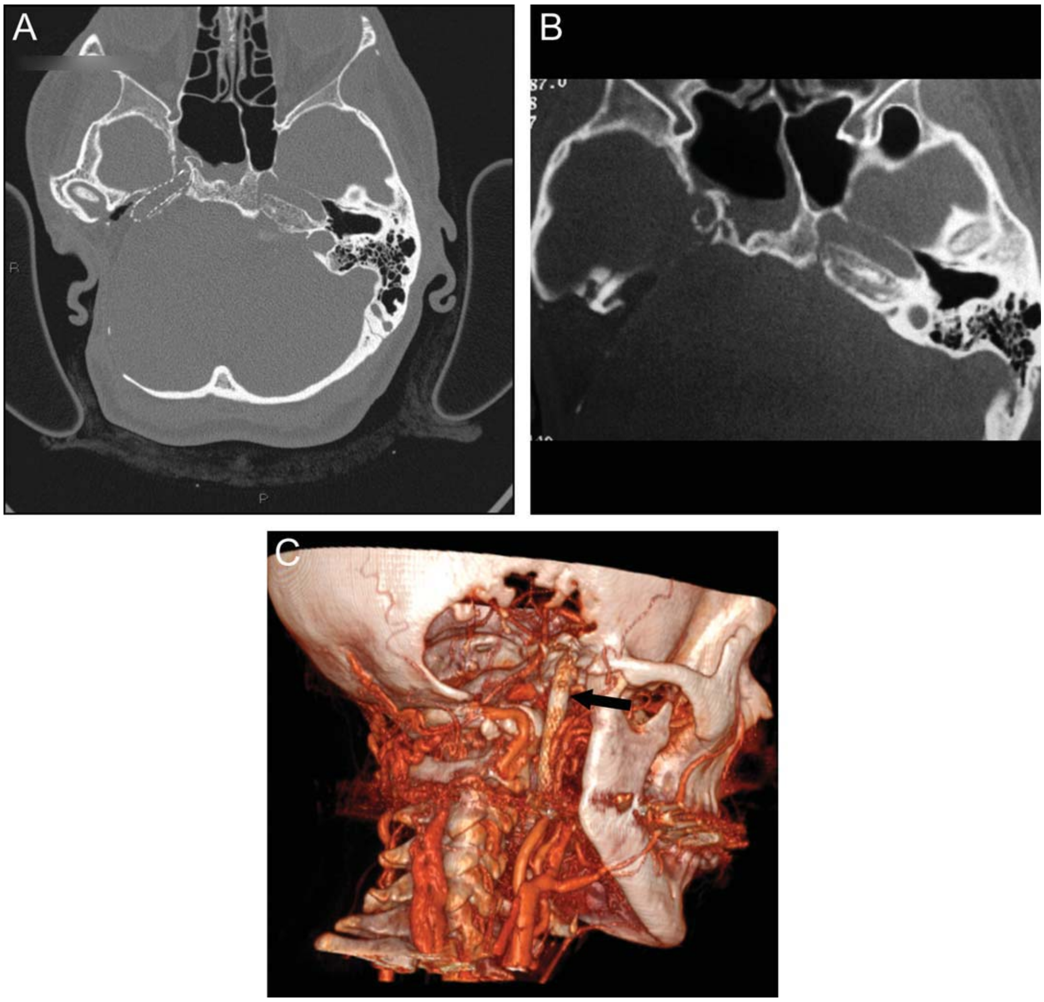

Fig. 3. The intraluminal stenting. (A) Axial computed tomography (CT) image showing the stent inserted into the horizontal ICA. (B) Axial CT image revealing the stent around the foramen lacerum. (C) Three-dimensional Angio-CT image demonstrating the amount of removed bone and the stent (black arrow) in the complex anatomical structures.

possible bilateral deficits in the LCNs, the importance of neural preservation can never be overemphasized. During first-stage surgery, the surgeon should pay attention to both tumor removal and LCN preservation.[6,39]

Our decision-making process in cases with bilateral or multiple paragangliomas is presented. In patients with LCN deficit on the side of a larger tumor, it is removed first and then the smaller tumor can be either followed-up or irradiated. On the contrary, if the patients have LCN deficit on the side of a smaller tumor, it is removed first and then the larger tumor can be followed-up with MRI. During follow-up, if the larger tumor shows evidence of growth, it may be partially removed with the preservation of LCN function or irradiated.

In patients with no LCN deficit, wait and scan is desirable first. However, if the tumor shows growth, radiotherapy or subtotal removal of the tumor with LCN preservation is performed first. Subsequently, if the tumor continues to grow in spite of radiotherapy or surgical removal, the other remaining modality can be utilized.

To preserve the function of LCNs, we should preserve the medial wall of the jugular bulb when dissecting the tumor.[6,40]

IFTA-A includes neck dissection and the cervical segment of the vagus nerve is always identified. In cases with vagal paraganglioma and TJP on the same side, simultaneous resection of both lesions is preferred.

We had 11 cases with multiple paragangliomas; five with TJP and ipsilateral VP; three with TJP and ipsilateral CBT; one with TJP and bilateral CBT; one with TJP, ipsilateral VP, and bilateral CBT; and one with TJP and contralateral CBT. Three were bilateral tumors.

Recurrence After Previous Surgery, Radiotherapy, or Stereotactic Radiosurgery

Residual or recurrent tumor masses can always be present when dealing with infiltrative skull base lesions. Complete radiographic reassessment is required to determine whether wait and scan, radiotherapy, or revision surgery is the optimal management. Any revision surgery is more difficult to perform when there are no normal tissue planes and surgical landmarks. Previous surgery or radiation increases the risk of CSF leak and damage to the LCNs and facial nerve.[4,10,11,13] Radiation tends to make tumor tissue firm, making its dissection and removal difficult.[13]

The carotid canal is the most common site for recurrence in TJP, and previous dissection increases the risk to the injury of the ICA. The importance of preoperative management of the ICA is especially highlighted as mentioned above. An IFTA-A with facial nerve rerouting should be performed in all cases with the appropriate extension. There is no place for a conservative approach for the facial nerve and external auditory canal in revision surgery.

In our present series, 13 cases had undergone previous treatment. Eight of them had various mastoidectomies without facial nerve rerouting, and the residual tumor was identified under the facial nerve and around the carotid artery during surgery. This was probably because facial nerve rerouting was not performed or there was incomplete tumor removal around the ICA. In certain cases, balloon occlusion or an intraluminal stent of the ICA is necessary in order to accomplish total tumor removal. To prevent residual tumor, extensive bone removal is essential due to the infiltrative characteristics of paragangliomas, and preservation of tumor-involved LCNs should not be attempted in young patients. In this study, the rate (76.7%) of gross total removal was lower than our previous report (90.7%)[1] due to the complexity of the cases.

In our opinion and based on our experience and data, surgical extirpation is the best choice for cure. However, some authors have insisted that radiotherapy has high tumor control rate and low morbidity and is as good an option for treating TJPs.[41–43] However, we noted that they grouped all paragangliomas together and did not analyze results for only TJPs. Also, they did not utilize any of the recognized tumor classifications, such as the Fisch classification, and had insufficient long-term follow-up. Therefore, it is difficult to directly compare the outcomes of surgery with gamma knife and radiosurgery as well as the morbidities as we cannot compare details such as neck extension and also intracranial extension. Our series had a large number of cases with intracranial extension although it is not clearly mentioned in any of the quoted articles. Additionally, the LCN morbidity in the aticles is comparable with our series even though our series had a large number of intracranial extensions.

CONCLUSION

Our experience in the management of complex cases of TJP has led us to a number of conclusions. IFTA-A is the cornerstone of this kind of surgery. Anterior rerouting of facial nerve should be performed to improve the control of the carotid artery and to prevent recurrence. The hemodynamics of the brain must be carefully investigated in the planning stage. In cases with bilateral tumors, LCN function should be preserved at least unilaterally to avoid compromising the patient's life. A proper preoperative endovascular intervention facilitates total removal. In cases where there is a large intradural component, staged removal is recommended to reduce the risk of CSF leak. Careful consideration of complicating factors and thorough preoperative evaluation and intervention can decrease surgical morbidity with a high probability of gross total removal. Certain tumor extensions remain unresectable without unacceptable morbidity, such as those that extend into the cavernous sinus, those encasing posterior circulation vasculature, or those with parenchymal brain invasion.

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