

# Absence of the Contralateral Internal Carotid Artery: A Challenge for Management of Ipsilateral Glomus Jugulare and Glomus Vagale Tumors

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**Objective:** Extensive involvement of the internal carotid artery (ICA) by jugular and vagal paragangliomas represents a challenging problem for skull-base surgeons: radical tumor resection and preservation of the vessel often cannot be achieved in these cases. The aim of this article was to report the management of a complex case of right ipsilateral jugular and vagal paragangliomas in a patient with absence of the contralateral ICA due to a previous removal of a left carotid body tumor. Surgical removal of the lesions was performed after stenting of the cervical and intratemporal segment of the ICA.

**Methods:** The charts and all the examinations of the patient were retrospectively reviewed. The patient, a 40-year-old man, was treated at the Gruppo Otologico, a private quaternary referral center for neurotology and skull-base surgery.

**Results:** The tumors were completely resected during surgery and manipulation of the ICA was greatly facilitated by the stent. No complication occurred during or immediately after surgery; after a follow-up period of 24 months there was no sign of recurrence and imaging studies demonstrated patency of the stented vessel.

**Conclusion:** This report suggests that placement of endovascular stents in the cervical and temporal segments of the ICA has the potential of changing the whole therapeutic management in cases of temporal and vagal paragangliomas that surround and invade the vessel, especially in those cases in which permanent balloon occlusion of the ICA is not feasible.

**Key Words:** Paragangliomas, stent, internal carotid artery.

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## INTRODUCTION

Jugular and vagal paragangliomas are generally benign, slow-growing neoplasms, associated with low morbidity and mortality. However, they have the potential to invade surrounding neurovascular structures at the skull base and extend intracranially; moreover, malignancy occurs approximately in 5% of jugular and 10% to 15% of vagal paragangliomas.<sup>1</sup>

Extensive involvement of the internal carotid artery (ICA) by jugular and vagal paragangliomas represents a challenging problem for skull-base surgeons: radical tumor resection and preservation of the vessel often cannot be achieved in these cases.

Although all the authors agree that complete excision offers the greatest opportunity for cure with acceptable morbidity in relatively simple cases, management of the ICA in complex cases has become an important matter of debate in the last years. It is difficult to decide between surgery and radiotherapy and, when surgery is preferred, it is a matter of discussion how extensive the resection should be and whether surgery should be preceded by any form of interventional endovascular procedure.

As recently reported,<sup>2</sup> the choice of the method of ICA management is based on accurate preoperative evaluation using imaging studies (high-resolution computed tomography [HRCT], magnetic resonance imaging [MRI], and 4-vessels digital subtraction angiography [DSA]) with the aim of clearly identifying the degree of the arterial wall involvement by the tumor and accurately visualizing the collateral cerebral circulation and the potential for permanent carotid occlusion. In the case of adequate collateral perfusion via the circle of Willis, permanent balloon occlusion is the simplest and most effective measure to manage an extensively infiltrated ICA. Stenting of the ICA represents an effective method of management of the vessel in cases of extensive arterial wall encasement by the tumor in the presence of inadequate collateral blood flow; reinforcing the arterial wall with the placement of a stent facilitates surgical dissection in a subadventitial plane and reduces the risk of vessel rupture during surgery.

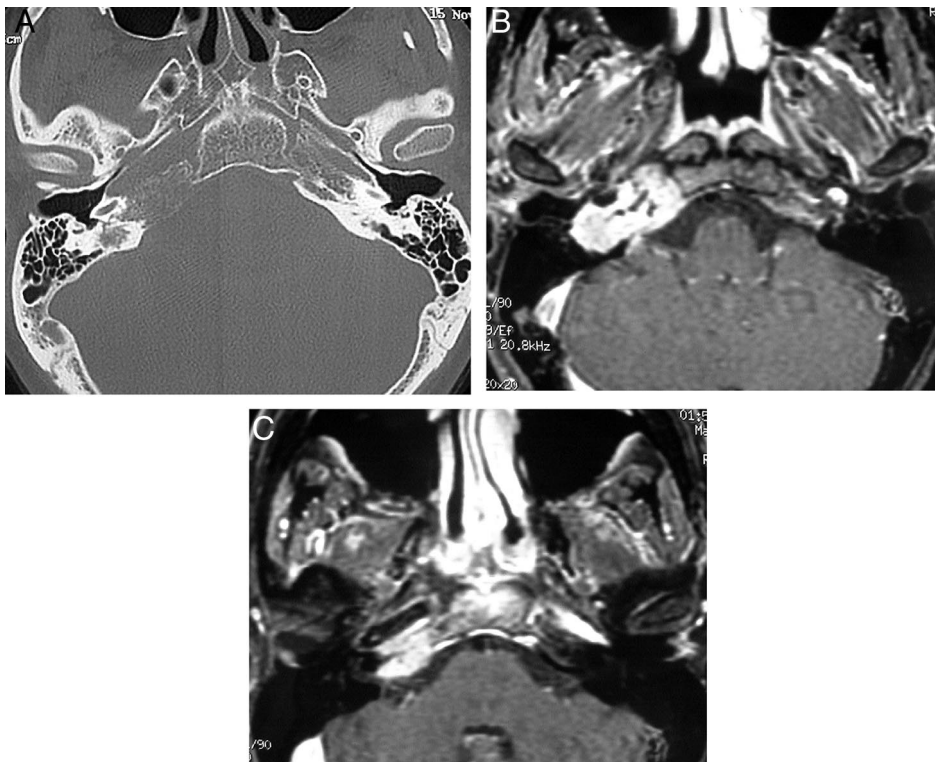


Fig. 1. High-resolution computed tomography scans showing bone erosion of the horizontal segment of the right internal carotid artery (ICA). (A) Gd-enhanced fat-suppressed magnetic resonance imaging showing tumor encasement of the vertical segment (B) extending along the horizontal segment (C) of the right petrous ICA. Occlusion of the left petrous ICA can be appreciated.

The aim of this article was to report the management of the ICA in a patient affected by two right ipsilateral paragangliomas (jugular and vagal) with extensive involvement of the vessel. The patient had been treated elsewhere 15 years before for a left carotid body tumor; during the surgical procedure the left common and ICAs had been ligated.

Surgical removal was achieved with subadventitial dissection after placement of an endoluminal stent with the aim of reinforcing the arterial wall. The challenging management and long-term results are reported.

## CASE REPORT

A 40-year-old man was referred to the Gruppo Otorico, a private quaternary referral center for neurotology and skull-base surgery, for a recently diagnosed right-sided jugular and vagal paragangliomas. Diagnosis was achieved by MRI of the head and neck, because of dysphonia that progressively worsened in a 3-year period.

The patient had been surgically treated elsewhere 15 years before for a left carotid body paraganglioma; during the previous procedure he had ligation of the left common and ICA.

On admission he complained of a right pulsating tinnitus and dysphonia.

The clinical examination showed a reddish mass behind the inferior aspect of an intact right tympanic membrane; the tongue appeared hypotrophic with fasciculation on the right side and the right soft palate and posterior pharyngeal wall were immobile. Flexible fiberoptic laryngoscopy showed palsy of the right vocal fold in a paramedian position with good compensation from the opposite side.

These findings indicated deficit of the lower right cranial nerves 9 through 12.

On the left side of the neck there was evidence of the scar of the previous transcervical surgical approach; left otoscopy and left side lower cranial nerves function were normal.

Audiometric threshold was bilaterally 20 dB for air conduction; facial nerve function was normal on both sides (grade I according to the House-Brackmann facial nerve function scale).

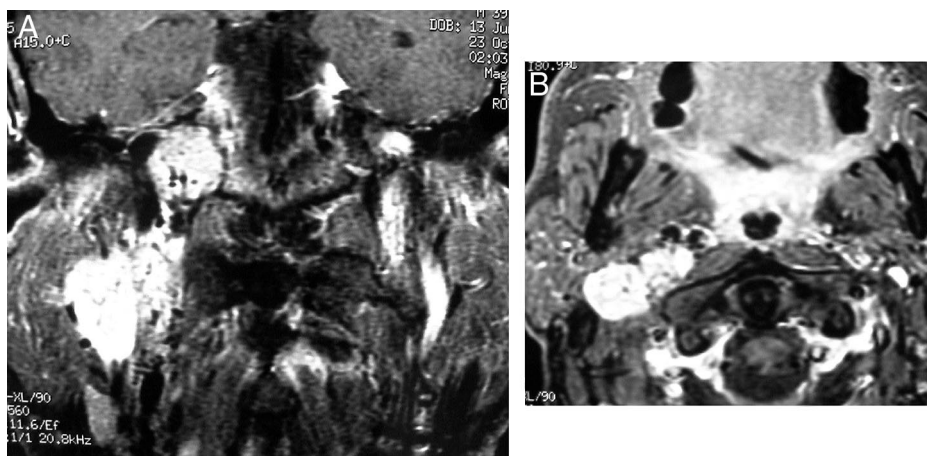
Evaluation was completed by imaging studies; the patient underwent HRCT of the temporal bone and skull base and four-vessel DSA. MRI scans performed elsewhere were accurately reviewed.

The computed tomography scan showed bone erosion of the vertical portion and along the medial wall of the horizontal portion of the petrous segment of the ICA. MRI showed two distinct masses at the level of the parapharyngeal space and jugular foramen, with encasement of the vertical segment of the ICA and extension along the horizontal segment of the ICA (Figs. 1 and 2). Angiography showed occlusion of the left common and ICAs; the left external carotid artery was retrogradely filled by a vertebro-occipital anastomosis. On DSA the lesions on the right side appeared at the level of the jugular foramen and the parapharyngeal space with classic tumor blush and venous drainage into the right internal jugular vein; the left internal jugular vein was occluded with hypertrophy of the left suboccipital venous plexus. There was no widening of the carotid bifurcation and both the internal and external carotid arteries were displaced anteriorly. The distal cervical segment and the vertical portion of the petrous segment of the right ICA were irregular and stenotic due to tumor encasement (Fig. 3). The flow compensation of the left cerebral hemisphere was from both the vertebrobasilar circulation and contralateral carotid artery.

With the help of imaging studies, tumors were classified as type I vagal paraganglioma and C3 De1 temporal paragangliomas.<sup>3</sup>

According to our previous experience<sup>2</sup> the patient underwent stenting of the right ICA with the aim of reinforcing the arterial wall; two Xpert stents, 6 mm in diameter and 40 mm in

Fig. 2. Magnetic resonance imaging after Gd enhancement in coronal (A) and axial (B) planes. In the coronal scan two distinct masses are evident at the level of the jugular foramen and parapharyngeal space. In the axial scan the relationship among the parapharyngeal vagal paraganglioma, the internal carotid artery, and the vertebral artery are shown.



length (Abbot Laboratories Vascular Enterprises, Abbot Park, IL) were inserted to cover the petrous and cervical segments of the right ICA (Fig. 4). The patient was given antiplatelet therapy starting 1 week before the procedure with the following scheme: acetylsalicylic acid 100 mg QD indefinitely and ticlopidine 250 mg BID for 1 month. Seven weeks after stenting, the patient underwent embolization with polyvinyl alcohol particles of the right ascending pharyngeal and occipital arteries.

Two days later he was operated on by the senior author (M.S.) through a right infratemporal fossa type A approach<sup>3,4</sup> and gross total removal was achieved. The ICA was freed from the tumor masses by creating a subadventitial plane of cleavage; having the ICA wall reinforced by the stents allowed for safe manipulation of the artery to control its whole circumference and reduced the risk of injury of the vessel. The operative field remained almost bloodless.

The postoperative period was uneventful and the patient was discharged 5 days after surgery.

The immediate postoperative right facial nerve function was grade VI according to House-Brackmann facial nerve function scale and there was a complete deficit of the right 9 through 12 cranial nerves.

Doppler ultrasound at discharge and at 6 and 18 months after surgery showed a patent right ICA.

Two years after surgery the patient's facial function had recovered to grade III and the right vocal cord palsy was well tolerated thanks to the contralateral compensation.

Contrast-enhanced MRI and HRCT at 6 and 18 months after surgery demonstrated patency of the right ICA and no signs of tumor recurrence.

## DISCUSSION

Debate exists in the literature regarding the appropriate treatment of the ICA in complex cases of paragangliomas.

A plan for management of the ICA in patients affected by jugular paragangliomas when the artery is encased and infiltrated in its cervical and intratemporal tracts can be accomplished after an accurate preoperative evaluation, aiming at estimation of the degree of tumor extension and at functional evaluation of the cerebral

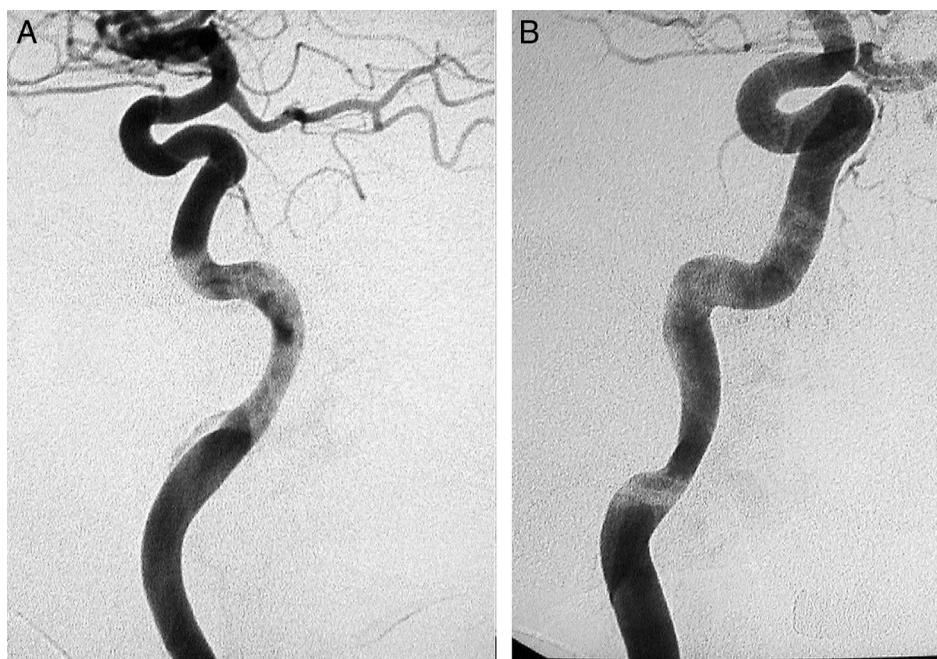


Fig. 3. Digital subtraction angiography of the right internal carotid artery in lateral (A) and oblique (B) views, showing stenosis of the distal cervical C1 segment and slight vascular supply from branches of the C4 segment.

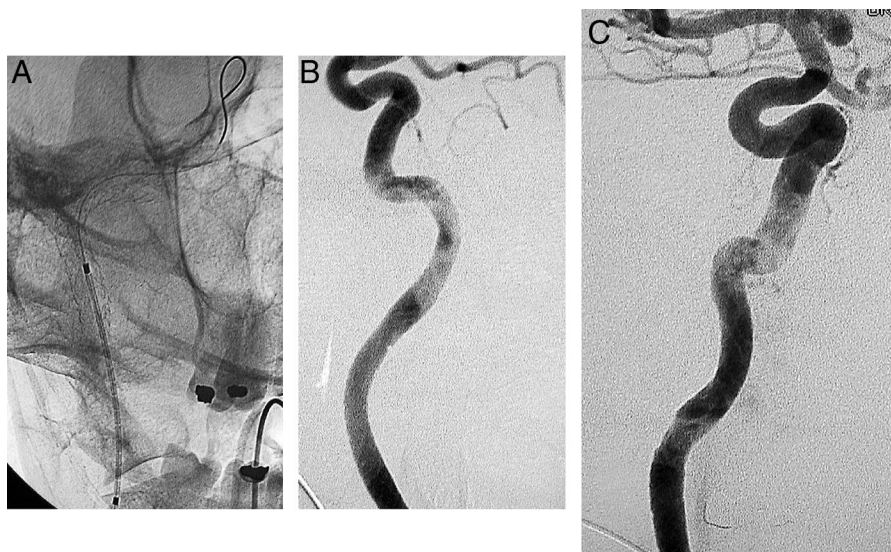


Fig. 4. (A) Plain X-ray showing a phase of the insertion of the stent in the C1 segment after a stent in the C2 segment has already been deployed. Notice partial overlapping of the stents. Lateral (B) and oblique (C) angiographic views of the internal carotid artery after stenting showing resolution of the stenosis.

blood flow. Adequate collateral flow through the circle of Willis permits permanent occlusion of the vessel using the balloon technique and the subsequent en bloc resection of the tumor and infiltrated vessel.<sup>5,6</sup> In the case of inadequate flow from the collateral circulation, clinicians may decide to wait and see, or choose for a by-pass procedure; both these procedures are not risk-free.

A first group of authors adopt a conservative management: Elshaikh et al.<sup>7</sup> believe that tumors that are so large that surgery would require ligating or reconstructing the carotid artery should be considered for radiotherapy to avoid possible surgical morbidity; Hinerman et al.<sup>1</sup> recommend primary resection for small-volume neck tumors; for large tumors morbidity to vascular structures has been shown if resection is attempted and radiotherapy alone is recommended in these situations.

In both cases the incidence of treatment-related acute complications was low, but long-term complications such as osteoradionecrosis, atherosclerosis, hearing loss, and development of second malignancies remain to be studied.

A second group of physicians have a more “aggressive” behavior claiming for a surgical removal without any form of preoperative endovascular neuroradiologic treatment of the ICA.

Netterville et al.<sup>8</sup> state that complete excision of vagal paragangliomas offers the greatest opportunity for cure with acceptable long-term morbidity and should be attempted in the majority of cases. He, however, recognizes that in some cases anatomic dissection of the ICA from the tumor can be extremely difficult and the surgeon must be prepared to suture lacerations during tumor excision and even to sacrifice or replace the ICA. He encountered problems with ICA in 5 of 46 cases and had one case of death and two cases of cerebrovascular accidents.

Miller et al.<sup>9</sup> noticed that despite a relative large size of his 19 cases of vagal paragangliomas, carotid artery injury did not occur with either the lateral cervical or midline mandibulotomy approach for surgical exposure.

Borba and Al-Mefty<sup>10</sup> stated that total removal of vagal paragangliomas must be attempted in all cases with

minimal morbidity and preservation of the ICA. He treated four cases of vagal paragangliomas and noticed that if the supraadventitial dissection plane is used, no permanent occlusion of the ICA is needed.

Patel et al.<sup>11</sup> state that major arterial encasement by tumor certainly deserves close scrutiny before surgical management. In patients with partial encasement of the ICA, dissection of the tumor is easier, whereas in patients with total encasement, repair of the ICA with temporary occlusion can be required. In his series of 12 patients, he encountered vascular problems in five cases (three ICA have been repaired and two were resected) without any complication. However, he concludes that these patients are certainly at greater risk for stroke complications.

Thabet and Kotob<sup>12</sup> experienced vascular injuries in two cases with one case of massive postoperative cerebral infarction, which resulted in death of the patient.

Billier et al.<sup>13</sup> found that invasion of the carotid wall in three patients with malignant intravagal paragangliomas required sacrifice of the carotid artery in each instance; he did not encounter complications.

A third group of physicians state that surgical removal of the lesions should be preceded by some form of preoperative endovascular neuroradiologic treatment of the ICA.

Zane et al.<sup>14</sup> described the largest series of balloon occlusion of the ICA in temporal paragangliomas. He treated 31 patients and noticed two minor and two major neurologic complications; one of the major complications occurred after intraoperative balloon occlusion and the other one was attributed to early surgery after preoperative balloon occlusion. He concludes that preoperative balloon occlusion can be safely performed in selected patients and that surgery should be delayed for a period of at least 1 to 3 weeks after carotid occlusion.

Sanna et al.<sup>15</sup> described seven patients (six temporal and one vagal paragangliomas) treated with surgery after preoperative balloon occlusion without any complications.

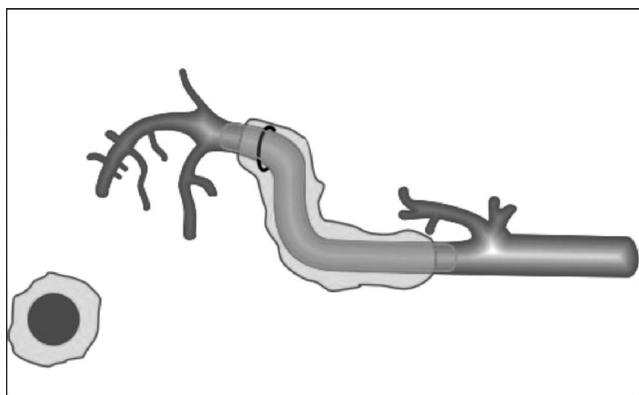


Fig. 5. Schematic drawing showing the stent reinforcement of the internal carotid artery.

Browne et al.<sup>16</sup> described 15 patients affected by vagal paraganglioma, nine of whom have been operated on after preoperative balloon occlusion of the ICA without complications.

More recently, after Cohen et al.<sup>17</sup> described a case report of temporal paraganglioma operated on after the insertion of a covered stent into the petrous carotid artery, Sanna<sup>2</sup> reported about three cases of temporal paragangliomas, which have been considered poor candidates for preoperative carotid occlusions, and were operated on with subadventitial dissection after insertion of nitinol stents into the petrous and cervical segments of the ICA, with good surgical results and absence of cerebrovascular complications.

In our Center we recently introduced stenting of the ICA in the surgical practice; as reported, surgical dissection was exceedingly easy with an almost bloodless surgical field<sup>1</sup> (Fig. 5). Having the arterial wall reinforced by the stent permits a safe subadventitial dissection and easy manipulation of the artery; the surgeon is in fact allowed to control the whole circumference of the vessel. The main drawback of placing a stent is the need for a lifelong antiplatelet therapy (i.e., acetylsalicylic acid 100 mg QD) with the potential risks associated. The main risks associated with the stenting procedure are distal embolization and thrombosis<sup>18</sup>; none of this occurred in our experience.

In the reported case, decision toward surgery has been taken after all the options had been evaluated together with the patient himself, considering the young age, the progressive worsening of the symptoms, and the already-present deficit of the lower cranial nerves 9 to 12. Accounting the previous left carotid artery occlusion and extensive right ICA involvement by the tumor, surgery had to be performed as safely as possible; having the arterial wall reinforced by the stent permitted a safe subadventitial dissection and easy manipulation of the ICA.

## CONCLUSIONS

In conclusion, we can state that placement of endovascular stents in the cervical and temporal segments of the ICA has the potential of changing the whole therapeutic management in cases of temporal and vagal paragangliomas, especially in those cases in which permanent

balloon occlusion of the ICA is not feasible; nevertheless a careful selection of patients is mandatory to minimize endovascular and surgical complications. At the present time, in our experience, irradiation should be reserved to treat small residual tumor after surgery and to control tumor progression in elderly patients and in patients with preexistent contralateral cranial nerve deficits.

It is a matter of fact that the incidence of cerebrovascular complications has been greatly reduced after the introduction of permanent balloon occlusion and stenting of the ICA in selected patients.

## BIBLIOGRAPHY

1. Hinerman RW, Mendenhall WM, Amdur RJ, Stringer SP, Antonelli PJ, Cassisi NJ. Definitive radiotherapy in the management of chemodectomas arising in the temporal bone, carotid body and glomus vagale. *Head Neck* 2001;3:363–371.
2. Sanna M, Khrais T, Menozzi R, Piazza P. Surgical removal of jugular paragangliomas after stenting of the intratemporal internal carotid artery: a preliminary report. *Laryngoscope* 2006;116:742–746.
3. Sanna M, Saleh E, Russo A, Taibah A. *Atlas of Temporal Bone and Lateral Skull Base Surgery*. Stuttgart/New York: Georg Thieme Verlag; 1995.
4. Sanna M, Russo A, De Donato G, Caruso A, Taibah AK. *Color Atlas of Otolaryngology*. Stuttgart/New York: Georg Thieme Verlag; 2002.
5. Andrews JC, Valavanis A, Fisch U. Management of the internal carotid artery in surgery of the skull base. *Laryngoscope* 1989;99:1224–1229.
6. Al-Mefty O, Teixeira A. Complex tumors of the glomus jugulare: criteria, treatment, and outcome. *J Neurosurg* 2002; 7:1356–1366.
7. Elshaikh MA, Mahmoud-Ahmed AS, Kinney SE, Wood BG, Lee JH, Barnett GH, Suh JH. Recurrent head and neck chemodectomas: a comparison of surgical and radiotherapeutic results. *Int J Radiat Oncol Biol Phys* 2002;2:953–956.
8. Nettekville JL, Jackson CG, Miller FR, Wanamaker JR, Glasscock ME. Vagal paraganglioma: a review of 46 patients treated during a 20 year period. *Arch Otolaryngol Head Neck Surg* 1998;124:1133–1140.
9. Miller RB, Boon MS, Atkins JP, Lowry LD. Vagal paragangliomas: the Jefferson experience. *Otolaryngol Head Neck Surg* 2000;122:482–487.
10. Borba LAB, Al-Mefty O. Intravagal paragangliomas: report of four cases. *Neurosurgery* 1996;38:569–575.
11. 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.
12. Thabet MH, Kotob H. Cervical paragangliomas: diagnosis, management and complications. *J Laryngol Otol* 2001;115: 467–474.
13. Biller HF, Lawson W, Som P, Rosenfeld R. Glomus vagale tumors. *Ann Otol Rhinol Laryngol* 1989;8:21–26.
14. Zane RS, Aeschbacher P, Moll C, Fisch U. Carotid occlusion without reconstruction: a safe surgical option in selected patients. *Am J Otol* 1995;16:353–358.
15. Sanna M, Piazza P, Di Trapani G, Agarwal M. Management of the internal carotid artery in tumors of the lateral skull base: preoperative permanent balloon occlusion without reconstruction. *Otol Neurotol* 2004;25:998–1005.
16. Browne J, Fisch U, Valavanis A. Surgical therapy of glomus vagale tumors. *Skull Base Surg* 1993;3:182–192.
17. Cohen JE, Ferrario A, Ceratto R, Miranda C, Lylyk P. Covered stent as an innovative tool for tumor devascularization and endovascular arterial reconstruction. *Neurol Res* 2003;25:169–172.
18. Assadian A, Senekowitsch C, Rotter R, Zolss C, Strassegger J, Hagmuller GW. Long-term results of covered stent repair of internal carotid artery dissections. *J Vasc Surg* 2004;40:484–487.