Head and Neck Paragangliomas: An Update on Evaluation and Management

Michael G. Moore, MD1, James L. Netterville, MD2, William M. Mendenhall, MD3, Brandon Isaacson, MD4, and Brian Nussenbaum, MD5

Abstract
Objective. Head and neck paragangliomas are a group of slow-growing hypervascular tumors associated with the paraganglion system. The approach to evaluate and treat these lesions has evolved over the last 2 decades. While radical surgery had been the traditional approach, improvements in diagnostic imaging as well as radiation therapy techniques have led to an emphasis on observation and nonsurgical therapy in many patients. This article reviews the contemporary approach to the workup and management of head and neck paragangliomas.

Data Source. Articles were identified from PubMed.

Review Methods. PubMed searches with the following keywords were performed: carotid body paraganglioma management, vagal paraganglioma management, jugulotympanic paraganglioma management, imaging of head and neck paragangliomas, head and neck paraganglioma embolization, paraganglioma radiation, head and neck paraganglioma management, observation of head and neck paragangliomas, bilateral carotid body paragangliomas, and genetics of paragangliomas. Review and original research articles available in the English language and published during or after 2009 were selected on the basis of their clinical relevance and scientific strength. Certain articles published prior to 2009 were also included if they provided background information that was relevant.

Conclusions/Implications for Practice. Workup and treatment of head and neck paragangliomas are changing. With more now known regarding the longitudinal behavior of these tumors, observation and nonsurgical therapy are indicated in many instances. For patients where surgery is the most appropriate option, improved diagnostic and perioperative techniques are allowing patients to tolerate resection, often with reduced morbidity.

Keywords
paraganglioma, carotid body paraganglioma, vagal paraganglioma, jugulare paraganglioma

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Paragangliomas are hypervascular tumors that can arise in the various locations of the paraganglion system. They are usually benign and slow growing, and approximately 90% of tumors occur in the adrenal paraganglia (so-called pheochromocytomas). In terms of extra-adrenal tumors, 85% occur in the abdomen, 12% in the thorax, and 3% in the head and neck. Carotid body paragangliomas (CBPs) are the most common in the head and neck, followed by jugular foramen and vagal tumors. Tumors in the larynx, nasal cavity, orbit, trachea, aortic body, lung, and mediastinum have also been described.

Traditional management of head and neck paragangliomas involved surgical extirpation. Due to the rich vascularity of these tumors, as well as their close association with neurovascular structures and/or the skull base of the infratemporal fossa, such procedures were often accompanied by significant blood loss, speech and swallowing dysfunction, and risks of stroke or even death. A number of innovations over the past few decades have improved the clinician’s ability to evaluate, counsel, and treat patients with these tumors and their families. This review focuses on these developments, ranging from improved diagnostic

1Department of Otolaryngology–Head and Neck Surgery, Indiana University School of Medicine, Indianapolis, Indiana, USA
2Department of Otolaryngology–Head and Neck Surgery, Vanderbilt University Medical Center, Nashville, Tennessee, USA
3Department of Radiation Oncology, University of Florida School of Medicine, Gainesville, Florida, USA
4Department of Otolaryngology–Head and Neck Surgery, University of Texas Southwestern Medical Center, Dallas, Texas, USA
5Department of Otolaryngology–Head and Neck Surgery, Washington University School of Medicine, St. Louis, Missouri, USA

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Corresponding Author:
Michael G. Moore, MD, Department of Otolaryngology–Head and Neck Surgery, Indiana University School of Medicine, 550 North University Blvd, Rm 3170, Indianapolis, IN 46202, USA.
Email: mgmoore@gmail.com
imaging and preoperative embolization techniques to nonsurgical strategies such as observation and treatment with stereotactic radiation therapy (RT). In addition, it provides an update on the genetics of paragangliomas, as well as strategies for managing bilateral tumors.

Methods

As a literature review, this study did not require Institutional Review Board approval. A PubMed query was performed with the following sets of words: carotid body paraganglioma management, vagal paraganglioma (VP) management, jugulotympanic paraganglioma management, imaging head neck paraganglioma, head neck paraganglioma embolization, head neck paraganglioma radiation, head neck paraganglioma management, observation head neck paraganglioma, bilateral head neck paragangliomas, and paraganglioma genetics. Review and original research articles available in the English language and published during or after 2009 were evaluated. Manuscripts were selected on the basis of their clinical relevance and scientific strength, per our discretion. Certain articles published prior to 2009 were also included if they provided background information that was relevant. Using this information as a guide, we provide clinical insight on these areas. In addition, we include a brief history of the evaluation and management of head and neck paragangliomas to put into context how these new findings have and will affect the treatment of patients in the future.

Discussion

Carotid Body Paragangliomas

CBPs are the most common head and neck paraganglioma. While first described in Europe in 1862 by Von Luschka, the first successful CBP removal in the United States was done by Scudder in 1903. Women are slightly more often affected than men, and the average age of diagnosis is 45 years. Due to their location away from the skull base, they are the most accessible head and neck paraganglioma for surgical removal. However, because of their intimate association with the carotid vessels and adjacent cranial nerves (CNs), careful evaluation and preparation prior to intervention are critical to ensure that risks of stroke and cranial neuropathies are minimized.

CBPs typically present as a slow-growing, painless lateral neck mass. In advanced disease, hoarseness or dysphagia may be present due to the involvement of the vagus nerve. Pulsatile tinnitus may also be appreciated on the side of the tumor due to the high-flow state. Physical examination often demonstrates a pulsatile lateral neck mass that is characteristically less mobile in the cephalocaudal direction due to adherence to the carotid artery. CN palsy (usually the vagus nerve) is present in up to 10% of patients. As a result, preoperative laryngoscopy is recommended to assess vocal fold motion.5

Imaging for CBPs should include magnetic resonance angiography (MRA) or computed tomography (CT) angiography. With use of either modality, an enhancing soft tissue tumor is characteristically seen at the level of the carotid bifurcation, splaying the internal and external carotid systems to create the “lyre sign” (Figure 1). Magnetic resonance imaging will demonstrate the classic “salt and pepper” pattern within the tumor, representing flow voids of its prominent blood vessels. Positron emission tomography, when combined with CT technology, has been shown to be an effective way of screening for multifocal paragangliomas.6,7

Following physical examination and imaging, CBPs can be stratified with the Shamblin classification system, as described in 1971. Based on this assessment, removal of advanced tumors comes at a higher risk of permanent CN functional loss.8,9 Such prognostication is relevant when the appropriate management strategy is being chosen. Options include observation, surgical excision, or RT.

For smaller tumors and for younger, relatively healthy patients, complete tumor excision is considered the treatment of choice, as this has been shown to result in local control in 94% to 100% of patients.9,11 However, complication rates vary significantly based on tumor size and vessel involvement. In fact, Papaspyrou and colleagues found that removal of tumors <5 cm resulted in a CN deficit rate of 14%, compared to 67% in patients with tumors >5 cm.12 When you consider that observation of paragangliomas has been shown to result in new or progressive CN deficits in 30% to 33% of patients13,14—a number similar to stereotactic radiosurgery (SRS)—CBPs <5 cm should be strongly considered for surgical removal. Additional factors, such as pain, concern for malignancy, or rapid growth, may further support operative intervention. Conversely, advanced age and associated medical comorbidities, as well as swallowing dysfunction, may make surgery less advisable.

For those ultimately managed with resection, careful preoperative planning and counseling are critical for a successful result, and collaboration with a vascular surgeon is recommended in nearly all cases.16 Tumor extirpation...
involves achieving adequate exposure, as well as proximal and distal vascular control. Associated CNs should be identified away from the lesion and dissected free, including the glossopharyngeal, vagus (and superior laryngeal nerve branch), spinal accessory, and hypoglossal nerves. Arterial dissection should next be performed, utilizing the periadventitial white line of Gordon from the “outside in,” leaving the central area of the carotid bifurcation until last. Figure 2 shows a transcervical exposure of a left-sided Shamblin class II tumor that has been mobilized from the carotid bulb. In the event that resection of the common or internal carotid artery wall is necessary, repair with a patch or interposition vein graft can be performed by a vascular surgeon.

Vagal Paragangliomas

VPs originate from 1 of the 3 ganglia of the vagus nerve, most frequently involving the nodose ganglion, and usually start within 2 cm of the vagal nerve’s extracranial course. They are much less common than their counterparts from the carotid body and jugular foramen, making up ≤5% of head and neck paragangliomas. As demonstrated by Netterville and colleagues, the most frequent presenting symptom is a neck mass, followed by hoarseness, with the vagus nerve being affected in approximately 28% of patients on presentation. The female:male ratio is 1.87:1, with the mean age at diagnosis being 43 years. They have also been shown to occur at a higher rate on the right side.

Evaluation starts with a detailed history and physical examination, including laryngoscopy to assess for vocal fold motion. CT can be helpful in evaluating the size of the tumor and its relationship to the anterior skull base. MRA is also helpful in determining the tumor’s blood supply and delineating the extent of surrounding soft tissue involvement. VPs tend to occupy the poststyloid parapharyngeal space and typically displace the carotid system anteromedially.

Management options for VPs are similar to those for carotid body and jugular foramen paragangliomas (JFPs). While operative intervention was traditionally the treatment of choice, due to the associated speech and swallowing morbidity following VP resection, it is now less commonly recommended. When necessary, surgery is usually accomplished through a lateral approach, starting with a transcervical preauricular approach to the infratemporal fossa with additional extension superiorly being provided via a trans-temporal approach, with or without facial nerve mobilization. Complete resection of these tumors usually requires removal of the associated vagus nerve, thus creating a unilateral vocal fold paralysis and pharyngeal plexus deficit, along with ipsilateral pharynx numbness and velopharyngeal insufficiency. In fact, in a series of 40 VPs, Netterville and colleagues found that the vagus nerve had to be resected in 37 patients and that all 40 patients developed a permanent vocal fold paralysis postoperatively.

Moreover, risks to other lower CNs range from 21% to 39% when a lateral approach is completed for resection of a VP or JFP.

In a meta-analysis by Suarez et al, data for 226 VPs undergoing resection revealed 93.3% local control with a mean follow-up of 86.7 months, a result similar to that seen following RT and SRS. However, more treatment-induced CN deficits were observed following surgery, compared with nonsurgical therapy. Therefore, for the majority of patients—especially the elderly, those with bilateral tumors, and when preexisting swallowing or pulmonary pathology exists—observation should be considered, with radiation being applied in cases of tumor progression.

Jugular Foramen Paragangliomas

As opposed to CBPs, JFPs are located at the lateral skull base and consequently pose a significant management challenge with respect to achieving tumor control with minimal morbidity. JFPs may present with a variety of symptoms,
including pulsatile tinnitus, hearing loss, dysphonia, shoulder weakness/pain, dysarthria, and facial paralysis. A complete head and neck physical examination readily identifies CN deficits and may allow the clinician to establish the diagnosis during otoscopy when a red retrotympanic mass is identified (Figure 4). JFPs typically present in the fifth and sixth decades of life and are 3 times more common in females than males. There is often a substantial delay (30.5 months) between onset of symptoms and diagnosis due to the slow growth rate of these tumors (0.8 mm/year).13

As with other head and neck paragangliomas, CT and magnetic resonance imaging (MRI) allow for differentiation of JFPs from tympanic paragangliomas as well as other temporal bone neoplasms. Irregular osseous erosion centered in the jugular foramen with further extension into the pneumatized spaces of the temporal bone is typically seen on CT. Magnetic resonance imaging also allows for determination of soft tissue and intracranial extension more readily than CT (Figure 5). Interventional angiography with embolization with or without internal carotid artery trial balloon occlusion can reduce intraoperative blood loss and provide information on intracranial collateral circulation prior to surgery.

Management options for JFPs are similar to those for CBPs and VPs and include observation, surgery, RT, or a combination therapy of surgical debulking, followed by radiation. Patient age, comorbidities, hearing status, CN function, and tumor size and location are just a few of the factors that dictate the approach to take. Observation is reasonable for many patients with JFPs, as the majority are benign and slow growing.

One concern in the management of JFPs is the progression or the development of new CN deficits. In one series, new lower CN deficits were noted in 30% of the patients undergoing observation.14 Carlson et al described the natural history in a series of 15 patients and 16 tumors where new-onset CN VII, X, XI, and XII deficits occurred in 6%, 13%, 13%, and 19% of patients, respectively. In this same study, progression of existing neuropathies involving CN X, XI, and XII occurred in 13%, 6%, and 6% of the patients undergoing observation, respectively. Moreover, 6 patients (38%) experienced progressive hearing loss, and 2 (12.5%) experienced bloody otorrhea from tumor extension into the external auditory canal.13

Surgical resection via a variety of approaches has been one of the mainstays of treatment for JFPs. Indications for resection include young age, secreting tumors, significant intracranial mass effect, tumor progression after radiation, facial paralysis, malignant transformation, and low risk of lower CN injury. Traditional surgical management through an infratemporal fossa approach entails overclosure of the external auditory canal and mobilization of the facial nerve, which results in a maximal conductive hearing loss and facial paresis. Mobilization of the facial nerve with the posterior belly of the digastric muscle and its investing fascia has substantially reduced the incidence of postoperative facial paresis.22 The fallopian bridge technique provides reduced access to the jugular foramen but essentially eliminates the risk of facial nerve paresis. The external auditory canal can be preserved in cases where there is minimal middle ear extension and limited involvement of the petrous carotid artery. A more conservative approach in the surgical management of JFPs has been recently proposed by several authors to reduce morbidity from injury to the lower CNs and decrease the risk of stroke and death.23-25

Figure 4. This otomicroscopic view demonstrates the view of a retrotympanic paraganglioma of the right ear.

Figure 5. This coronal T1-weighted contrast-enhanced image demonstrates a right-sided jugular foramen paraganglioma, showing the relationship of the tumor and the lateral skull base. An arrow points to a feeding vessel at the inferior aspect of the tumor.
For tumors with significant intracranial extension as well as involvement of the middle ear and mastoid, a combined approach has been described where tumor in the middle ear and mastoid is removed and the remaining jugular foramen and intracranial component is treated with radiation. Advantages of this approach include resolution of preoperative conductive hearing loss, reducing or eliminating pulsatile tinnitus, decreasing the risk of hemorrhage from the external auditory canal, reduced risk of lower CN injury, and reduction of the radiation dose to the cochlea.

A recent meta-analysis by Ivan et al. provided an in-depth examination of tumor control, morbidity, and mortality for JFP. Gross total tumor excision results in a tumor control rate of approximately 86% (81%-91%). Approximately 60% of patients experienced new lower CN deficits after gross total tumor excision. The incidence of CN IX, X, XI, and XII deficits after surgery were 38%, 26%, 40%, and 18%, respectively. In addition to lower CN deficits, facial nerve paresis, profound hearing loss, and mortality occurred in 35%, 45%, and 3%, respectively, after gross total excision.

RT for JFPs, like surgical therapy, has evolved over time with advances in technology. In fact, it has been shown to result in similar disease control to that of surgery but with a reduced risk of CN injury. In a separate meta-analysis by Ivan et al, mean tumor control was shown to be as high as 95% after RT. New CN IX, X, XI, and XII deficits were identified in 9.7%, 9.7%, 12%, and 8.7%, respectively. Chun and colleagues recently reported 100% tumor control in 30 patients with JFPs using a hypofractionated approach.

**Carotid Artery Management and Preoperative Embolization**

In patients undergoing surgical resection of head and neck paragangliomas where carotid artery resection and reconstruction are considered, it is critical to plan appropriately to avoid intraoperative catastrophe. As previously discussed, the patient’s age, prognosis, and comorbid status must be evaluated, and a vascular surgery consult should be obtained. In addition to standard preoperative imaging, carotid artery balloon occlusion testing with postocclusion single-photon emission computed tomography can be used to assess for adequacy of cerebral perfusion. While informative, false-negative rates of up to 10% have been observed, and patients should be counseled appropriately. Additionally, the contralateral carotid artery might be at risk in the future from a metachronous tumor or development of atherosclerotic disease. As a result, it is our preference to repair/reconstruct the common and internal carotid system, when possible. In such instances, intraoperative electroencephalography can be performed to monitor for any changes that could indicate ischemia, and the use of a shunting device (Figure 6) following adequate intraoperative anticoagulation can be implemented to further minimize the risk of stroke. Figure 7 shows a saphenous vein graft patch that was used to repair the carotid bulb and proximal internal carotid artery following resection of a Shamblin class III tumor.

For the majority of JF and high VPs, preoperative embolization is recommended due to difficulty that can be encountered in obtaining proximal and distal vascular control prior to tumor manipulation. Despite the benefit that comes with reducing blood loss and operative times for these tumors, significant side effects have been encountered, such as stroke and cranial neuropathies.
conflicting data, however, concerning the benefit of embolization in CBPs. Power et al, in their series of 144 tumors, found that preoperative embolization reduced intraoperative blood loss by more than a half but resulted in no drop in operative time or temporary CN injury.\textsuperscript{35} Little and colleagues found that the use of embolization prior to tumor removal had no impact on operative time, complications, blood loss, or hospital stay for tumors up to 5 cm.\textsuperscript{36} In addition, while the risk of isolated cranial neuropathies is theoretically lower following embolization of CBPs, they have been reported.\textsuperscript{37} When embolization is deemed appropriate, it should be performed 24 to 48 hours prior to surgery to minimize revascularization as well as local edema and inflammation.

**Observation of Head and Neck Paragangliomas**

As outlined in the above sections, while treatment of cervical paragangliomas with surgery and/or RT yields the best likelihood of eradicating/controlling disease, these approaches often come with significant short- and long-term morbidity. As a result, the concept of disease observation has become more palatable to patients and physicians.

This approach has been supported by numerous recently published series. Langerman et al presented their group of 47 tumors in 43 patients, observed over a mean of 5 years. There were 28 CBPs and 19 VPs. During the study period, 42% remained stable, 38% grew, and 20% regressed. Those that enlarged did so at a mean growth rate of 2 mm per year.\textsuperscript{38} The same institution recently published its series of 16 JFPs observed over a mean of 86 months. It found that 52% remained stable while 48% grew at a mean rate of 0.8 mm/year.\textsuperscript{13} In a similar study by Prasad et al, 47 JFPs were managed with the “wait and scan” approach, and 79% remained stable or regressed during the period of observation. Those with a longer follow-up were more prone to show disease progression.\textsuperscript{14} New or worsening cranial neuropathies have been shown to develop in 30% to 33% of patients undergoing observation,\textsuperscript{13,14} a rate comparable to that of patients treated with SRS.\textsuperscript{15}

Based on these findings, the option of close observation should be considered for elderly patients, those with significant comorbidities, and those who are asymptomatic or minimally symptomatic. For those being observed, if possible, MRI is recommended to avoid regular exposure to radiation. While there are no validated protocols for serial imaging, we prefer to reimage in 6 months (symptomatic patients) or 12 months (asymptomatic patients), with an additional annual assessment for stable lesions. Indications for earlier imaging and/or treatment include concern for malignancy, progression of symptoms/cranial neuropathies, significant tumor growth, or pain.

**Management of Head and Neck Paragangliomas with RT**

As previously discussed, paragangliomas of the head and neck may also be managed with RT or SRS. In patients with larger tumors, particularly those arising in the skull base, surgical removal may require sacrifice of ≥1 CNs resulting in permanent long-term morbidity. RT\textsuperscript{39,40} and SRS\textsuperscript{41} are effective, safe treatment alternatives for such patients.

SRS is suitable for head and neck paragangliomas that are <3 cm in maximum dimension. An attractive feature of SRS is that it is administered in 1 fraction and is thus convenient. The goal is to administer a single dose of ablative irradiation (12.5-15 Gy) to the paraganglioma with a very steep dose falloff so that the adjacent normal tissues receive minimal radiation. Investigators at the Mayo Clinic reported on 42 patients (19 for primary treatment and 23 for recurrent tumors) with a mean tumor volume of 13.2 cm\textsuperscript{3} treated with Gamma Knife SRS and a mean margin dose of 14.9 Gy.\textsuperscript{42} Progression-free survival was 100% at 3 and 7 years and 75% at 10 years. Thirty-one percent of tumors decreased in size, 67% remained stable, and 2% grew. Fifteen percent of patients developed new CN deficits. Facial paralysis and deafness were more common in patients with recurrent tumors than in those receiving primary treatment (48% vs 11%, \(P = .02\)).

RT is an excellent treatment alternative for patients who are unsuitable for surgery or SRS.\textsuperscript{43} The likelihood of tumor control is as good or better than surgery or SRS, and the risk of severe complications, including irradiation induced malignancy, is <1%. The disadvantage of RT is that it requires a 5-week course of treatment. Radiation is administered with an external beam usually employing 6-MV x-rays and an intensity-modulated radiotherapy technique with a 5-mm margin. The RT dose is moderate (45 Gy in 25 fractions over 5 weeks) and very unlikely to cause significant morbidity. Gilbo et al recently reported the outcomes of 131 patients with 156 benign head and neck paragangliomas treated with RT with a median follow-up of 8.7 years.\textsuperscript{40} The 10-year outcomes were as follows: local control (lack of progression), 96%; cause-specific survival, 97%; and overall survival, 72%. No patient experienced a severe complication.

**Management of Bilateral Tumors**

In the case of bilateral head and neck paragangliomas, additional considerations apply. In these individuals, often there is an underlying genetic predisposition, thus putting them at higher risk of developing additional lesions. As a result, evaluation by medical genetics is indicated. Additional factors to consider include prior neck surgery or RT, the patient’s baseline neurologic function and life expectancy, as well as swallowing function and pulmonary reserve. For elderly patients or those with preexisting dysphagia or lung pathology, observation or RT may be more appropriate. Moreover, tumor factors such as size, type, and location influence decision making. For instance, surgical resection would be contraindicated if bilateral vagale tumors were present.

In individuals where surgery is considered, it is necessary to do it in a staged fashion to minimize the risk of bilateral cranial neuropathies and/or impact on cerebral circulation. The choice of which side to do first is a matter of debate,
and there is no conclusive literature to guide clinicians. We make this determination on the basis of perceived risk to the adjacent CNs. For individuals where 1 tumor is large, the smaller side is first excised to be sure that this can be accomplished without associated nerve injury and dysphagia. If difficulty is encountered, the options of observation or RT for the remaining tumor should be strongly considered. For individuals where both tumors are small to medium, the larger side is usually addressed first since the smaller tumor could be observed or radiated if nerve injury occurs at the time of the first operation.

An additional issue that plagues patients with bilateral CBPs is the potential for hemodynamic instability due to the loss of the carotid baroreceptor feedback system. In patients having undergone surgery for bilateral CBPs, they can develop significant hypertension from baroreflex failure due to the loss of innervation from both carotid sinuses/bodies. These individuals may also suffer hypotension when they are put under sedation in the future. Perioperative management consists of sodium nitroprusside to avoid hypertension. For more long-term control, clonidine (a peripheral and central alpha-2 agonist) or phenoxybenzamine (an alpha-1 and alpha-2 antagonist) may be used. In such instances, observation or RT is often strongly considered for the second side in an effort to avoid these issues.

**Genetics of Head and Neck Paragangliomas**

While the majority of paragangliomas are solitary, in approximately 10% of affected individuals, multiple paragangliomas are observed (usually bilateral CBPs). For patients with multiple tumors, there is approximately a 30% to 50% risk of a familial syndrome (usually autosomal dominant inheritance), and it is recommended that the extended family of patients with multiple tumors be screened with MRI. In such instances, genetic counseling is becoming increasingly recommended to detect tumors early and allow for treatment with fewer side effects.

In addition to succinate dehydrogenase mutations, patients with sporadic head and neck paragangliomas can have mutations in any one of at least 5 other gene sites, with an overall incidence of genetic abnormality nearing 50%. In addition to sporadic occurrence, these tumors can occur as part of multiple endocrine neoplasia syndromes (ie, types IIA and IIB) as well as neurofibromatosis and von Hippel-Lindau disease. In type IIA (Sipple’s syndrome), a germline mutation in the RET proto-oncogene on chromosome 10 results in a triad of medullary thyroid carcinoma, pheochromocytoma, and parathyroid hyperplasia. Type IIB also involves the RET gene but at a different site, and it has the additional component of mucosal neuromas. As a result, for patients with multifocal disease, family members with paragangliomas, early onset of their tumors, or VPs, genetic screening for mutations may be appropriate.

**Implications for Practice**

As described so far, the approach to management of head and neck paragangliomas is in evolution. Evaluation is similar for all lesions, with up-front history and physical examination being performed along with CT and/or MRI/MRA. For patients with multifocal tumors and those with a family history of similar or other tumors suggestive of a familial syndrome, medical genetics referral is recommended. At-risk family members can then be screened to identify tumors early to minimize morbidity of treatment.

It has been increasingly recognized that up-front nonsurgical management is often appropriate for head and neck paragangliomas. In all cases, it is recommended that patients be presented at a multidisciplinary tumor conference attended by participants representing head and neck surgery, neurology, neurosurgery, radiation oncology, pathology, speech and swallowing pathology, and neuroradiology. With the lack of available data to develop a concrete algorithm for management, we consider not only the chronicologic age of the patient but also his or her associated medical comorbidities, prior treatment, likelihood of developing additional lesions, and preexisting swallowing dysfunction. A thoughtful discussion should then be held between the clinician and the patient to determine the most appropriate management strategy.

Instances where surgery should be considered include young healthy patients with small to medium CBPs
(Shamblin class I or II or those <5 cm) and select JFPs where removal has a low risk of morbidity. Additional instances where surgery should be considered include secreting tumors, when there is a concern for malignancy, or when additional radiation cannot be offered. For the majority of other tumors, up-front observation is appropriate, with repeat imaging and office evaluation being performed in 6 to 12 months, depending on symptoms. For progression of symptoms or tumor growth, SRS or RT should be considered. By following this approach, unnecessary morbidity can be avoided while still providing acceptable tumor control.

Moving forward, prospective study is needed to further predict the clinical behavior of these neoplasms. As we gain additional insight into which tumors are likely to progress, recur, or be multifocal, we will be able to better counsel our patients and their families on the best management strategy to optimize disease control while minimizing associated morbidity.

Author Contributions

Michael G. Moore, conception, acquisition of data, analysis and interpretation of data, drafting and revising, final approval; James L. Netterville, analysis and interpretation of data, drafting and revising, final approval; William M. Mendenhall, analysis and interpretation of data, drafting and revising, final approval; Brandon Isaacson, analysis and interpretation of data, drafting and revising, final approval; Brian Nussenbaum, conception, acquisition of data, interpretation of data, drafting and revising, final approval.

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