Operative Techniques in Otolaryngology (2016) 27, 25–29

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Operative Techniques in Otolaryngology

Surgical management of vagal paraganglioma

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KEYWORDS
Vagal paraganglioma;
Parapharyngeal space tumor;
Surgical management of vagal paraganglioma;
Postoperative complications

Vagal paragangliomas are nonchromaffin tumors that originate from paraganglionic chemoreceptor cell bodies associated with the vagus nerve. Although both surgical excision and radiation therapy have been reported to be effective treatment modalities, the effectiveness of one over the other remains unclear. There are several surgical approaches that have been described. As with any surgical disease, the utility of a technique over another has been justified at some point in time. Clearly, there has been a shift away from the more radical approaches with the primary goal of surgery being complete extirpation of the tumor along with avoidance of iatrogenic cranial neuropathies and preservation and restoration of cranial nerve function. In addition, early rehabilitation of speech and swallowing dysfunction results in improved functional outcomes and reduced hospital stay and morbidity.
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Introduction

Vagal paraganglioma (VPG) or glomus vagale is a rare neoplasm representing fewer than 5% of all head and neck paraganglioma. They are characteristically slow growing benign tumors with low morbidity. However, they can often be variable in their clinical aggressiveness because of their potential to invade surrounding neurovascular structures. Surgical excision of these tumors is advocated as the definitive treatment. Alternatively radiation therapy has been advocated. The relative efficacy of one modality over the other remains unclear. In a recent metaanalysis, Suarez et al,[1] reported a 93.3% long-term control of the disease in 226 VPG with surgery. A comparison of surgery with radiation therapy could not be done because of limited numbers. In many instances, watchful waiting is considered as a reasonable option that avoids the morbidity from either treatment modality.

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http://dx.doi.org/10.1016/j.otot.2015.12.007
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Clinical presentation

VPG most often remain asymptomatic for a long time. They may not present with any specific signs or symptoms and are only diagnosed incidentally on head and neck imaging for an unrelated condition. However, when present, the most common presentation is a neck mass, which may be present in almost half the patients. Other clinical signs and symptoms may include pulsatile tinnitus, pharyngeal swelling, or dysphonia. Less commonly, dysphagia, facial weakness, conductive hearing loss, aural fullness, cervical bruit, Horner syndrome, or headache may be present.

Radiographic evaluation

Radiological assessment is necessary not only for diagnosing VPG, but also to plan appropriate surgical treatment. Both computed tomography (CT) with contrast and magnetic resonance imaging with gadolinium provide information necessary to fulfill both these objectives. Of

the 2, magnetic resonance imaging is the more informative study. The tumor typically appears hyper intense on both T1 and T2 weighted images and enhance with gadolinium. The classic "salt and pepper" appearance is obvious in many cases because of low signal intensity flow voids, produced by high vascular flow.

Bilateral carotid angiography provides information about involvement of the carotid artery and also provides opportunity for conducting a balloon occlusion test when the carotid artery may be involved with the tumor and carotid resection may potentially be required. It also provides the option of performing selective preoperative embolization of this highly vascular tumor.

Patient selection

The rationale of surgical excision of VPG is based on certain patient factors, tumor characteristics, and the availability of multimodality specialty services that should include head and neck surgery, neurootology, neurosurgery, and vascular surgery. Equally important consideration must be given to the potential complications of cranial nerve (CN) dysfunction resulting from not just the loss of function of CN X, but also from the potential injury or sacrifice of CNs IX-XII.

Surgery should not be considered in all patients. Elderly patients, those with high-risk comorbidities, patients with intact CN function, asymptomatic patients with small tumors, or patients with bilateral tumors are best monitored for tumor growth and CN dysfunction. Additionally, if there is contralateral lower CN paralysis or paresis, then surgery may not be a suitable option because of significant bilateral cranial neuropathies. In these cases, radiation therapy may be a reasonable option if there is evidence of progression of disease.

Surgical anatomy

VPG are mostly confined to the neck and parapharyngeal space (PPS), with only the more advanced cases involving the jugular foramen and lateral skull base, and even more rarely, extending intracranially. The importance of knowing the anatomy of the PPS and the skull base cannot be over emphasized.

The PPS is an inverted pyramid shaped area that starts at the skull base and extends caudally to its apex at the greater cornu of the hyoid bone (Figure 1).

The lateral walls of the nasopharynx and oropharynx limit the medial extent of the PPS. These are formed by the superior constrictor muscle and the pterygomandibular raphe and lined by the buccopharyngeal fascia. The PPS tumors tend to grow out of the confines of the space in a medial or inferior direction because of lack of fascial barriers or bone.[2]

The lateral boundary is formed by the fascia on the medial surface of the parotid gland and by the pterygoid muscles.

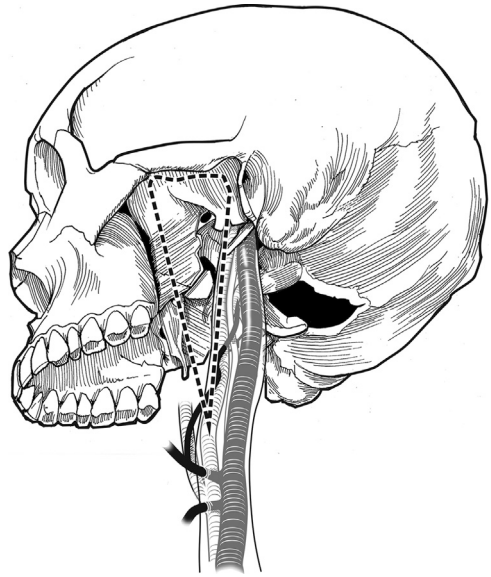

Figure 1 The parapharyngeal space is an inverted pyramid with its base at the skull base.

The prevertebral muscles form the posterior wall. However, as the carotid sheath is a separate deep neck space, it may be considered as the posterior wall of the PPS.

The superior boundary is the skull base underlying the middle cranial fossa. The infratemporal fossa may be considered as a direct superior extension of the PPS.

Surgical approaches to the VPG

The surgical approaches and extent depends on the location of the tumor. Netterville et al,[2] described this eloquently based on 3 possible extensions of the tumor. The tumor may be confined to the cervical region or the PPS. From the cervical region, it may extend to the lateral skull base up to the jugular foramen and cause anterior and medial displacement or even encasement of the internal carotid artery (ICA). Finally, in more advanced cases, it may extend into the jugular foramen often with intracranial extension. Hence, the surgical approach must be considered in a step-wise progression starting from a cervical route, extending laterally along the mastoid, exposing the jugular foramen, and ultimately exposing the skull base for tumors extending medially or intracranially.

Transcervical approach

A transverse cervical incision preferably in a prominent skin crease is used for making the incision a couple of fingerbreadths below the inferior border of the mandible. Subplatysmal flaps are raised. The marginal mandibular nerve is preserved. This nerve is located in the superficial layer of the deep cervical fascia and is most easily identified at the angle of the mandible. The submandibular gland is generally preserved, but it may be removed to obtain a broader exposure for larger tumors (Figure 2). Cervical

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