Acquired Vascular Tumors of the Head and Neck

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KEYWORDS

• Paraganglioma • Carotid body tumor • Glomus jugulare tumor • Glomus vagale
• Juvenile nasopharyngeal angiofibroma • Hemangiopericytoma

KEY POINTS

- Acquired head and neck vascular tumors are rare and account for less than 5% of head and neck neoplasms.
- Management of head and neck paragangliomas has evolved from primarily surgical to more conservative treatment consisting of observation and nonsurgical therapy.
- The mainstay treatment of juvenile nasopharyngeal angiofibroma and hemangiopericytoma is surgery, with radiation reserved for adjuvant therapy and recurrent tumors.

INTRODUCTION

Vascular tumors of the head and neck consist of a diverse group of both benign and malignant neoplasms, many of which arise in close association with blood vessels or tissues. Head and neck vascular tumors are exceedingly rare, with each type accounting for no more than 0.5% of head and neck neoplasms. Although varying greatly in biologic behavior, with exceptions of a few, most of these tumors are similar in their indolent growth and tendency for local recurrence.

Vascular neoplasms of the head and neck present with a wide spectrum of signs and symptoms. Patients usually complain of nonspecific symptoms, which often have been present for a prolonged period of time. Diagnosis, therefore, requires a high index of suspicion and is usually made after these tumors are large enough to be visually apparent or cause symptoms. This article discusses the most common acquired benign and malignant vascular tumors, with an emphasis on their evaluation and treatment.

PARAGANGLIOMA

Natural History and Physical Findings

Paragangliomas are vascular neoplasms that arise from the extraadrenal paraganglia derived from the neural crest and most commonly occur in the head and neck region.

Disclosure: The authors have nothing to disclose.
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Otolaryngol Clin N Am ■ (2017) ■–■
https://doi.org/10.1016/j.otc.2017.09.015 **oto.theclinics.com**

These tumors are closely associated with either blood vessels (carotid artery, jugular bulb) or nerves (vagus, tympanic plexus). Paragangliomas are usually slow-growing tumors with an average growth rate of 1 to 2 mm per year and a median doubling time of 4.2 years. Their growth pattern may be described as biphasic because very small and very large paragangliomas have a lower growth rate when compared with that of intermediate-sized tumors.[1]

Although all paragangliomas have the potential of releasing vasoactive substances, such as catecholamines and dopamine,[2] only 1% to 3% of paragangliomas produce the associated clinical findings.[2,3] Secreting paragangliomas release norepinephrine, and a 4-fold to 5-fold elevation of serum norepinephrine is necessary to produce symptoms,[4] such as excessive sweating, hypertension, tachycardia, nervousness and weight loss.[2] Urinary laboratory screening tests, including 24-hour urinary metanephrine (normal <1.3 mg) and vanillylmandelic acid levels (normal range is 1.8–7.0 mg), are frequently elevated 10 to 15 times normal in patients with actively secreting tumors.[2] Serum catecholamine levels, including norepinephrine and epinephrine, are also of value in the evaluation of the patient.

Multicentric and hereditary paragangliomas

A familial history of paragangliomas may be present and there is a significant incidence of multicentric tumors in both familial and sporadic cases. Familial or hereditary paragangliomas have been previously reported to account for 5% to 10%[5,6] of all cases of head and neck paragangliomas but it seems that these estimates were low due to the complex mode of inheritance and variable phenotypic expression.[7] It may, in fact, account for up to 25% to 50% of cases.[8,9] Most (90%) cases of hereditary paragangliomas involve the carotid body.[10] If a familial history is present, there is a 78% to 87% possibility of multiple paragangliomas.[6,11] Bilateral carotid body paragangliomas occur more frequently with familial cases (31.8%) than nonfamilial cases (4.4%).[5]

The potential to develop multicentric tumors has important clinical implications. The presence of bilateral carotid paragangliomas poses a difficult challenge in management because excision of these tumors results in loss of baroreceptive function and subsequent refractory hypertension. Multiple tumors, including vagal or jugular paragangliomas, present problems concerning significant morbidity of multiple lower cranial nerve (CN) dysfunction, perhaps bilaterally, resulting from direct tumor involvement or surgical resection. Because multicentric tumors may be metachronous, routine follow-up MRI, ^{111}indium pentetreotide (Octreoscan) or fluorine-18-labeled dihydroxyphenylalanine (^{18}F-DOPA) PET imaging is indicated.

Malignant variant

Malignant paragangliomas are uncommon and their diagnosis can only be confirmed by metastatic disease, usually within regional lymph nodes, because histologic examination of the primary tumor is unreliable for establishing a malignant diagnosis. The prevalence of malignancy depends on site of the primary tumor and there has been considerable variability in the reported frequency. Although malignant carotid body tumor has been reported in up to 20% of patients, most reports indicate a rate of 3% to 6% of cases.[12,13] Malignancy is generally less common in familial paragangliomas compared with sporadic cases.[5] The jugulotympanic paraganglioma malignancy rate ranges widely from less than 1% to 25% but is most often reported to be approximately 5%.[13,14] Vagal paragangliomas probably represent the highest rate of malignancy (16%–19%) of the more common types of head and neck paragangliomas.[13]