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Treatment of carotid paraganglioma

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Carotid paragangliomas, commonly known as carotid body tumors, are the most commonly reported head and neck paragangliomas. They are slow growing and always associated with the carotid arteries and adjacent nerves. They are rarely malignant. The estimates for familial transmission of this tumor range from 30%-50%. When multiple paragangliomas are present, the most common combination is 2 carotid paragangliomas. There are no absolute indications for surgical resection, but tumors expressing biochemical activity, a growth rate >1-2 mm per year or demonstrating airway or alimentary tract compression should be resected. Patients younger than 50 years should have small-to-medium sized paragangliomas resected. Patients with significant comorbidities or a contralateral cranial nerve dysfunction should not have surgery. Large carotid paragangliomas involving the skull base would likely result in multiple lower cranial nerve dysfunction after resection requiring multispecialty care afterwards. When dealing with bilateral carotid paragangliomas, resection of the smaller tumor first is recommended. Simultaneous carotid paraganglioma excision is discouraged because of acute baroreceptor failure. The second carotid paraganglioma may be observed for growth or it may be radiated depending on symptoms, size, or growth. The role and indications for angiography and embolization are covered elsewhere in this issue. If a carotid paraganglioma is biochemically active, alpha-adrenergic blockade followed by beta blockade is recommended and should be commenced 10 days before surgery. The complications associated with carotid paraganglioma excision are lower cranial nerve palsies and cerebrovascular accidents, although these are quite uncommon for small-to-medium sized tumors. Meticulous surgery and control of the carotid arteries can decrease these morbidities dramatically. Recently, a "top down" or craniocaudal approach to these tumors has gained acceptance among many surgeons. This approach controls the cephalad internal carotid artery and external carotid artery and removes the tumor down to the bifurcation. This technique has decreased the incidence of cranial neuropathy and blood loss in these patients. Following treatment, the uncomplicated patient requires yearly follow-up consisting of physical examination and magnetic resonance imaging. The familial patient should be genetically tested for succinate dehydrogenase mutations. Depending on the type of mutation yearly imaging and evaluation of siblings, parents and children is strongly advised. As metachronous lesions are possible, this follow-up is a lifetime undertaking.

Introduction

Before discussing the treatment of carotid paragangliomas, it is necessary to consider the biological behavior of these tumors. Classically, they are slow growing tumors that invade locally with a propensity for invasion or investment of local nerves and arteries. In a study of 26 patients,

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Jansen et al[1] determined the growth rate to be 0.83 mm per year for a group of patients with carotid paragangliomas. Overall, 29 lesions showed a volume increase and were considered to be enlarging. Additionally, death from the presence of nonmalignant paragangliomas is a rare event.

With this demonstrated tumor behavior, indications for carotid paraganglioma surgery can be complex decisions depending on the age of the patient, the size of the tumor, preoperative cranial nerve involvement, anticipated postoperative cranial nerve paralysis, patency of the circle of Willis to provide for cross-cerebral circulation, multiplicity of paragangliomas, and the involvement of local organs (esophagus, soft palate, and larynx). Available surgical and interventional vascular radiological expertise should also be taken into consideration. Evaluation of carotid circulation, tumor flow dynamics, displacement of the major vessels, and the prediction of tolerance of interruption of the internal carotid circulation must be ascertained before any undertaking.

Surprisingly, there are no accepted definite indications for carotid paraganglioma resection. The relative rarity of the tumor makes larger studies uncommon. The need for multidisciplinary approaches, both diagnostically and therapeutically, make firm guidelines difficult to tabulate.

There are some indications for carotid paraganglioma surgery that we would consider mandatory. Biochemically active tumors and tumors showing growth rates $>$1-2 mm per year would most likely dictate surgery. Tumors that compress the airway or alimentary tract resulting in functional breathing and swallowing problems or causing obstructive sleep apnea should be considered for surgery. Young patients with preexisting lower cranial neuropathy or vocal fold paresis or paralysis should undergo surgery. Patients diagnosed with familial paraganglioma syndrome, who demonstrate no evidence of cranial nerve dysfunction, should undergo resection of small paragangliomas if cranial nerve preservation is anticipated.[2]

In contradistinction, carotid paragangliomas that may not require resection include older patients with multiple comorbidities. If preexisting contralateral lower cranial nerve dysfunction is present, especially involving the vagus nerve, no operation should be considered. Carotid paraganglioma resection involving very large tumors involving the skull base may likely result in multiple lower cranial dysfunction and should be avoided. If the contralateral side has already been resected, some surgeons would elect observation of the second carotid paraganglioma to avoid baroreceptor failure and resulting significant labile hypertension. In this situation, continued observation of the carotid paraganglioma is warranted and, if necessary, external beam radiation therapy is highly effective in controlling the tumor.

The most common combination of multiple paragangliomas is bilateral carotid paragangliomas (Figure 1). This combination is most frequently found in familial paraganglioma syndrome patients. The phenotype usually presents earlier than sporadic paragangliomas. In these cases, we suggest resection of the smaller paraganglioma.

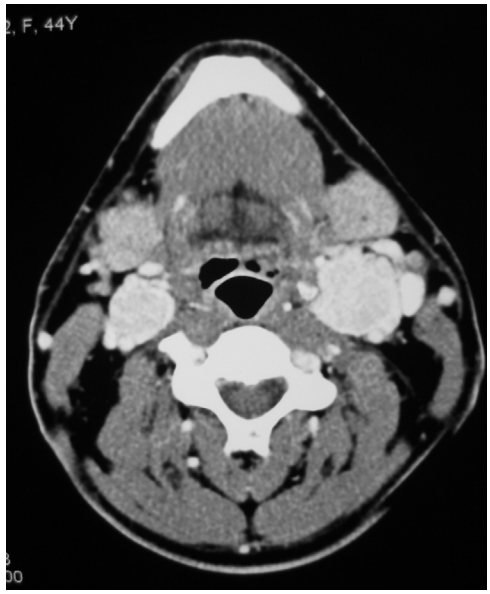

Figure 1 CT scan of bilateral carotid paragangliomas. (Color version of figure is available online.)

The second lesion can be observed, radiated, or resected if growth is observed.[3,4]

The details of preoperative evaluation of patients selected for operative intervention is covered elsewhere in this issue. If there is a history of hypertension, serum metanephrines should be evaluated. Serum levels of metanephrine have been found to be more specific, easier to collect, and not dependent on dietary intake as compared with 24-hour collected urinary levels.[5] Imaging either withcomputed tomography angiography or magnetic resonance imaging (MRI) would evaluate for local invasion, pathologic lymph nodes, vascular involvement, and skull base invasion. MRI is the preferred study because of the definition of local invasion, characteristic appearance of paragangliomas, the ability to evaluate vascular structures, and the lack of ionizing radiation (compared with computed tomography scanning). The characteristic "salt and pepper" appearance on MRI is diagnostic for paragangliomas. Some authors have recommended angiography with or without embolization, but there is no consensus of the necessity of this modality for carotid paragangliomas. Most experienced surgeons do not use embolization for carotid paragangliomas. There does not appear to be uniformity of opinions regarding embolization. The indications and technique are addressed elsewhere in this issue. Larger carotid paragangliomas are embolized by many experts.[6] Much of the role of angiography alone has been supplanted by less invasive MRI or magnetic resonance angiography and computed tomography angiography.

Shamblin et al[7] described a classification system to grade the extent of carotid paraganglioma involvement of the carotid system, which reflects the potential complexity of resection. Type I tumors are relatively small with minimal attachment to the carotid vessels. Type II carotid paragangliomas are larger with moderate arterial attachment, whereas a Shamblin type III represents complete encasement