Healthy 73-year-old woman was seen for a slowly enlarging right cervical mass located near the angle of her mandible. She had symptoms of fever, chills, and hoarseness. Physical examination demonstrated a firm, nontender, pulsatile mass without associated lymphadenopathy. A bruit was present over the mass on auscultation. A computed tomography scan was performed, showing a 3 × 2-cm mass located between the external and internal carotid arteries (ICA) that enhanced with intravenous contrast injection (Figure 1). The mass was partially wrapped around the ICA. There was no evidence of invasion into the surrounding tissues, nor was there a mass in the contralateral neck. A cerebral arteriogram showed “splaying” of the external carotid artery and the ICA, with blushing seen on delayed images (Figure 2).

What Is the Most Appropriate Management for This Patient?
A. Angiographic embolization of blood supply to the mass
B. Surgical resection of the mass
C. External beam radiation
D. Close observation with repeated computed tomography or magnetic resonance scans

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Carotid Body Tumor: Intercarotid Paraganglioma

Carotid body tumors are rare lesions of the carotid bifurcation (CB), arising from chemoreceptive tissue of the carotid body located on the posteromedial aspect of the CB. Neoplasms involving the carotid body are derived from mesodermal elements of the third arch and neural crest ectoderm. Occasionally referred to as intercarotid paraganglioma, they occur in 0.01% of the population; however, there is a 9.5% familial incidence.1,2 These tumors have a less than 10% incidence of local or distant metastasis. Malignancy is difficult to assess, as patients tend not to have recurrence for many years following resection of a primary tumor, and histological invasion of the tumor is not evidence of malignancy.

These tumors were originally classified by Shamblin et al3 according to the degree of involvement with the ICA. Tumors in group 1 have no arterial attachment and are resectable without significant trauma to the arterial wall or tumor capsule. Group 2 tumors partially surround the ICA, while those in group 3 completely circumscribe the ICA. Larger tumors also tend to be more adherent to the arterial wall.

The first line of investigation for a cervical mass that appears to be pulsatile should be duplex ultrasonography. This study will distinguish solid or cystic masses, identify carotid aneurysm or kinked carotid arteries, and demonstrate splaying of the CB by a mass, as in the case of a carotid body tumor.5,6 Computed tomography scanning is a useful tool for investigating the extent of invasion into an adjacent structure, particularly intracranial extension or pharyngeal involvement.6 Additional diagnostic studies will demonstrate splaying of the CB and tumor enhancement. Angiography will also rule out the possibility of bilateral tumors, identify the blood supply of the lesion, and assess the intracranial circulation.

The vascularity of these tumors raises the possibility of angiographic embolization. Embolization has been reported as a useful adjunctive preoperative treatment as it may reduce operative blood loss. In reality, the feeding vessels are small, difficult to localize, and selectively catheterize. The role and importance of radiation therapy in tumor management is uncertain. There is some evidence for local control with occasional tumor regression. However, the definition of local control is vague, as recurrence often occurs throughout several decades.

Surgical resection is the mainstay of therapy, as 10% of cases are malignant, with lymph node involvement or distant metastases being documented. If tumors are resectable, no adjuvant therapy is recommended. Close follow-up observation with interval computed tomography scanning is advisable. As with standard carotid surgery, care must be taken to avoid injury to neighboring cranial nerves (mainly nerves X and XII) that may have been displaced by the tumor mass (Figure 3).

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