Case report

Bilateral neck paragangliomas

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Summary

Paragangliomas of the head and neck are rare neoplasms presented as cervical mass, generally bilateral, that arise from chemoreceptors located at the carotid bifurcation (carotid body tumors), along the vagus nerve (vagal paragangliomas), and in the jugular fossa and tympanic cavity (jugulotympanic paragangliomas). They are typically asymptomatic at the beginning, highly vascular, slow-growing and compressing the surrounding anatomic structures. Only radical surgery is the curative treatment for paragangliomas. We present a case of a 62-year-old woman with a diagnosis of bilateral neck paragangliomas where surgical removal was judged burdened by excessive risk because of the size of the tumor.

A 62-year-old woman with a previous diagnosis of bilateral neck paragangliomas presented with a 3-month history of dry cough, hoarseness and dysphagia. Thirty-five years earlier, she had noticed a painless, slowly enlarging bilateral cervical mass. Since she was otherwise asymptomatic, she did not seek medical attention. Physical examination revealed a mobile, well-defined, hard bilateral mass of her neck (behind the angle of the mandible to right side) under the sternocleidomastoid muscle; the right side of her neck had more swelling than left (Figure 1A). A mild bruit was heard on masses auscultation. Inspection of the oral cavity showed medial bulging of the right pharyngeal wall and tonsil. Indirect laryngoscopy revealed a paralyzed right vocal cord. Screening for catecholamines and their metabolites were unremarkable. A Color Duplex flow image of lateral neck showed a bilateral hypervascular masses surrounding and filling the carotid bifurcation without any significant compression (right mass in Figure 1B). A contrast-enhanced computed tomography (CT) of the brain and the neck showed large soft-tissue bilateral carotid space mass with a homogenous and intense enhancement (Figure 1C). The 3D volume-rendering reconstructions showed the left-sided carotid body tumor (Figure 1D) causing marked widening of the carotid bifurcation. A coronal magnetic resonance imaging showed (Figure 1E) heterogeneous hypersignal of the tumor with the typical ‘salt and pepper’ image (arrowheads) corresponding to a highly vascularized mass and lateral pharyngeal wall bulging into the pharyngeal lumen (arrows). The 111-indium-labeled octreotide scintigraphy, used mainly to detect multicentric or metastatic paragangliomas was positive only in the regions of bilateral cervical ganglions (Figure 1F). Paragangliomas of the head and neck (carotid body tumors) are rare neoplasms presented as cervical mass, generally bilateral, that arise from chemoreceptors. They are typically asymptomatic at the beginning, highly vascular, slow growing and compressing the surrounding anatomic structures. In the head and neck area, most common sites, paragangliomas are

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Figure 1. (A) The patient with bilateral mass of her neck (behind the angle of the mandible to right side) under the sternocleidomastoid muscle; the right side of her neck has more swelling than left. (B) Color Duplex flow image of lateral neck showing a right hypervascular mass surrounding and filling the carotid bifurcation. (C) Contrast-enhanced CT of the brain and the neck showing large soft-tissue bilateral carotid space mass with a homogenous and intense enhancement. (D) Multislice spiral CT angiography with 3D volume-rendering reconstructions showing the left-sided carotid body tumor causing marked widening of the carotid bifurcation. (E) Coronal magnetic resonance imaging showed heterogeneous hypersignal of the tumor with the typical ‘salt and pepper’ image (arrowheads) corresponding to a highly vascularized mass and lateral pharyngeal wall bulging into the pharyngeal lumen (arrows). (F) The 111-indium-labeled octreotide scintigraphy was positive only in the regions of bilateral cervical ganglions.
mainly found in the carotid body, jugular foramen, the middle ear and vagal glomus. The curative treatment for paragangliomas is a radical surgery. Most paragangliomas are benign and, as a result, prognosis depends directly on the site of the tumor, the size it may reach and the possibility of completely removing the tumor, as well as possible surgery-related complications. Over the recent years, adjuvant techniques have been incorporated, such as embolization, in order to decrease intra-operative bleeding, make surgery easier and reduce complications. However, in our patient, surgical removal was judged burdened by excessive risk because of the size of the tumor. As surgery removal was not an option, embolization and radiation therapy may be the definitive treatment.

References