Undifferentiated metastatic carcinoma and myoepithelioma: Two rare causes of hypervascular masses of the parapharyngeal space

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Abstract
We report 2 unusual cases of hypervascular masses in the parapharyngeal space. The first case involved a poorly differentiated metastatic carcinoma of oropharyngeal origin that mimicked a carotid body tumor. The second case involved a highly vascular myoepithelioma located in the parapharyngeal space.

Introduction
The three most common tumors of the parapharyngeal space are salivary gland tumors, neurogenic tumors, and metastatic tumors. In this article, we describe 2 unusual cases of hypervascular masses in the parapharyngeal space.

The first case involved a poorly differentiated metastatic carcinoma of oropharyngeal origin that mimicked a carotid body tumor. Carotid body tumors are the most common paragangliomas found in the parapharyngeal space.1 Para-gangliomas are tumors of neural crest origin that are made up of chief and sustentacular cells surrounded by stroma2; they account for 0.6% of all neoplasms of the head and neck.1 A poorly differentiated carcinoma that mimics a carotid body tumor is rare.

The second case involved a highly vascular myoepithelioma located in the parapharyngeal space. Myoepithelioma was first reported by Sheldon in 1943.3 Since then, only about 200 cases have been reported in the literature. These tumors typically involve the major salivary glands; they are not a common cause of an oropharyngeal mass.

Case reports
Patient 1. A 56-year-old man presented with an asymptomatic right-sided neck mass. His medical history was significant only for treated hypertension. In view of the morphologic characteristics of the mass, fine-needle aspiration biopsy was not performed. Diagnostic angiography demonstrated a heterogenous, hypervascular mass immediately adjacent to the right carotid bifurcation. The mass was supplied by a proximal external carotid artery feeder. The origin of the feeder was close to the origins of the ascending pharyngeal artery and the occipital artery (figure 1, A).

The feeding artery was embolized with polyvinyl alcohol (PVA) and a 2 × 4-mm detachable fibrin coil. The tumor was excised the next day (figure 1, B), and the patient was discharged after a successful recovery. Surgical pathology identified the resected specimen as a poorly differentiated carcinoma, presumably of metastatic origin. Staining was positive for pan-cytokeratin and negative for chromogranins, nonspecific esterase, synaptophysin, and S-100, supporting the diagnosis.

Magnetic resonance imaging (MRI), positron-emission tomography (PET), and gastrointestinal, pulmonary, and urinary evaluations did not detect any primary cancer. Specifically directed biopsies of the head and neck revealed a few atypical squamous islands in the lingual tonsil and a moderately to poorly differentiated, focally keratinizing, and partly necrotic squamous cell carcinoma in the tonsillar tissue.

Patient 2. A 68-year-old man with a medical history significant for hypertension and chronic atrial fibrillation presented to his dentist complaining of escalating pain over his right mandible during the previous 2 weeks. A dental examination revealed a large mass protruding into the oropharynx and oral cavity. Computed tomography (CT) detected a large, right parapharyngeal mass with enhancement suggestive of hypervascularity (figure 2, A). Diagnostic angiography demonstrated a highly vascular
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Lesion arising from the sphenopalatine branches of the internal maxillary artery (figure 2, B).

Prior to surgical excision, embolization with 45 to 150-μm and 150 to 250-μm PVA particles achieved good tumor hemostasis (figure 2, C). The tumor was completely resected. Histopathology revealed that it was a minor salivary gland myoepithelioma (figure 3, A and B). The patient was discharged 4 days after surgery, and he continued to do well at the 6-month follow-up.

Discussion
Paragangliomas of the head and neck may arise at the bifurcation of the carotid artery, at the jugular foramen, along the vagus nerve, or in the middle ear. The classic presentation of the most common paraganglioma of the parapharyngeal space—the carotid body tumor—is a non-tender, asymptomatic, slowly growing neck mass at the bifurcation of the common carotid artery. On angiography, a carotid body tumor appears as a hypervascular mass at the carotid bifurcation; it is characterized by tumor blush and splaying of the internal and external carotid arteries. Typically, the pharyngeal branch of the external carotid artery supplies the mass.

CT and MRI findings are keys to the diagnosis, and they can be useful in differentiating a carotid body tumor from meningiomas or other neurogenic tumors. On contrast-enhanced CT, carotid body tumors are homogeneous with intense enhancement. On MRI, they demonstrate low...
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1, biopsies of the oropharyngeal territory were performed, often necessary to identify the primary lesion. However, MRI and PET did not reveal a primary cancer, nor was there any evidence of direct extension from the nasopharynx or metastasis to the parapharyngeal space. Carotid body tumors may displace the internal carotid artery anteriorly and medially. Therefore, MRI, and angiography. Carotid body tumors may displace the internal carotid artery posteriorly and laterally. Thus, they are differentiated from vagal paragangliomas, which may displace the internal carotid artery anteriorly and medially. 

Immunohistologic analysis is critical to establishing a diagnosis, particularly staining for chromogranins and S-100. Chromogranins are proteins within the chief cells, and S-100 is produced by sustentacular cells (SY38 is another, less specific, marker for neuroendocrine tumors). Positive staining for chromogranins and S-100 establishes a mass of myoepithelial cells. 

Upon excision, the hypervascular mass of our patient 1 did not stain positive for chromogranins or S-100, suggesting a different etiology. The absence of other staining characteristics and the tumor’s morphology confirmed the diagnosis of an undifferentiated carcinoma, presumably of metastatic origin. The tumor could have arisen as a result of direct extension from the nasopharynx or metastasis to the parapharyngeal space. However, MRI and PET did not reveal a primary cancer, nor was there any evidence of direct extension from the oropharyngeal area. 

Difficulty establishing the primary cancer is not uncommon in metastatic lesions that arise from oropharyngeal cancers. In fact, biopsies of the oropharyngeal region are often necessary to identify the primary lesion. In our patient 1, biopsies of the oropharyngeal territory were performed, and a moderately to poorly differentiated infiltrating squamous cell carcinoma was identified in the tonsil. 

Other investigators have reported that papillary carcinomas of the thyroid, a leiomyoma, and schwannomas (among others) have appeared as carotid body tumors, but we have not seen any report of an undifferentiated mass that mimicked a carotid body tumor. This case represents an unusual presentation, and it highlights the importance of splaying in establishing a diagnosis. 

Myoepithelial cells are the smooth-muscle cells that surround ducts. Tumors of myoepithelial cell origin are made up entirely or predominantly of myoepithelial cells. These tumors are rare, accounting for 0.3 to 1.5% of all salivary gland neoplasms. Approximately 48% of myoepitheliomas occur in the parotid gland, 42% occur in the minor salivary glands, and the remainder occur in the submandibular gland and the seromucinous glands of the nasal cavity and larynx. Myoepitheliomas can also occur in any structure that has ducts. They have also been reported in the breast, lung, and pancreas. 

Myoepitheliomas occur at most ages. The mean age of onset is approximately 40 years. It is suspected that the myoepithelial cells that cause these tumors are involved in inhibiting angiogenesis. This suspicion represents a possible explanation for why differentiated myoepithelial tumors should not be vascular lesions. Myoepitheliomas typically present as slowly enlarging masses. Some are asymptomatic, and some cause pain by compressing local structures. These tumors are typically well encapsulated or circumscribed. 

A parapharyngeal mass is most often caused by a pleomorphic adenoma of the deep parotid gland or minor salivary glands. The differential diagnosis of a parapharyngeal mass includes lymphadenopathy, a branchial cleft cyst, a salivary gland tumor, an aneurysm of the carotid artery, a neurogenic tumor, a pleomorphic adenoma, and a paraganglioma. A paraganglioma is a likely cause of a vascular lesion of the parapharyngeal neck space. In contrast, a myoepithelioma is a rare cause of a parapharyngeal mass, although in some cases it might mimic a vascular neck mass, such as a paraganglioma. 

The treatment of a myoepithelioma involves complete excision with tumor-free margins. These tumors usually behave in a benign fashion, but there exists a potential for malignant transformation that necessitates close follow-up. The reason for the neovascularity of the tumor in our patient 2 is unknown, but such a finding is important because the diagnosis of myoepithelioma should be considered in the differential diagnosis of a vascular parapharyngeal space neck mass.

References


