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Severe Dry Cough as Presenting Symptom of Patient with Five Paragangliomas

Emily Waddick[1], Ameya Asarkar[1], Mansour Mirfakhraee[2], Cherie-Ann O. Nathan[1,*]

[1]Department of Otolaryngology/Head Neck Surgery, LSU Health, Shreveport, LA
[2]Department of Radiology, Willis Knighton Medical Center, Shreveport, LA
*Corresponding author: cnatha@lsuhsc.edu

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Abstract *Introduction:* A 60 year-old white female presented with a dry cough of 8 months duration. *Case report:* On examination, there was a reddish mass at the inferior right tympanic membrane and bilateral upper neck fullness. Subsequent imaging revealed five paragangliomas - right glomus jugulare, left glomus vagale, left carotid body tumor, right glomus vagale, and left superior mediastinum paraganglioma. *Discussion:* This case highlights the presenting symptoms, multidisciplinary workup, and management of a rare case of five paragangliomas.

Keywords: *paraganglioma, parapharyngeal space tumors, radiation therapy*

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1. Introduction

A 60 year-old female presented with a dry cough of 8 months duration. On examination, there was a reddish mass at the inferior right tympanic membrane and bilateral upper neck fullness. Subsequent imaging revealed five paragangliomas - right glomus jugulare, left glomus vagale, left carotid body tumor, right glomus vagale, and left superior mediastinum paraganglioma. She was treated with low dose amitriptyline for neurogenic cough. This case highlights the atypical presenting symptoms, multidisciplinary workup, and management of a rare case of five paragangliomas.

2. Case Report

A 60 years-old white female developed a dry cough of 8 months duration, treated initially with conservative management by her primary care doctor with a low dose amitriptyline for neurogenic cough. A chest computed tomography (CT) scan was performed which noted a contrast enhancing mass of the superior mediastinum/lower neck and she was referred to ENT. On exam, she had a reddish mass at the inferior right tympanic membrane and bilateral upper neck fullness/mass. She denied any dysphagia or voice changes. During the workup, she began to note dull right ear pain. She had a history of acoustic trauma with known deafness in her right ear for 25 years. Cranial nerves appeared intact. Informed consent was obtained from the patient to present her case report and de-identified radiological images.

2.1. Investigations

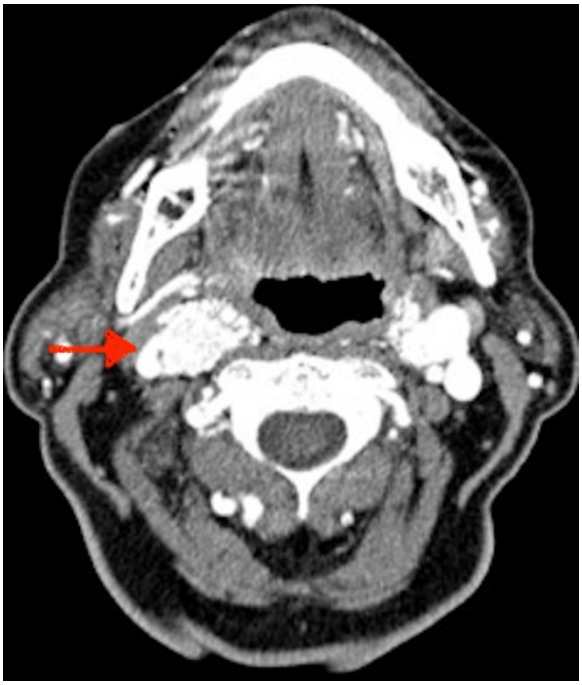

Figure 1. Axial CT neck with contrast showing right glomus vagale measuring 41x27mm extending to a point just above carotid bifurcation. Right internal carotid artery is displaced anteromedially. Also noted left glomus vagale (incompletely pictured)

Contrast enhanced Computed tomography (CT) scan of the neck and temporal bone was ordered as well as metanephrines. These scans showed five paragangliomas - right glomus jugulare, left glomus vagale, left carotid

body tumor, right glomus vagale, and left superior mediastinum paraganglioma. Axial CT neck with contrast showed right glomus vagale measuring 41x27mm extending to a point just above carotid bifurcation. (Figure 1). Axial CT temporal bone without contrast showed right glomus jugulare with classic irregular erosion and "moth eaten" appearance to temporal bone with extension to lower right CP angle and extending superolaterally towards hypotympanic region with erosive changes in the jugular bulb (Figure 2).

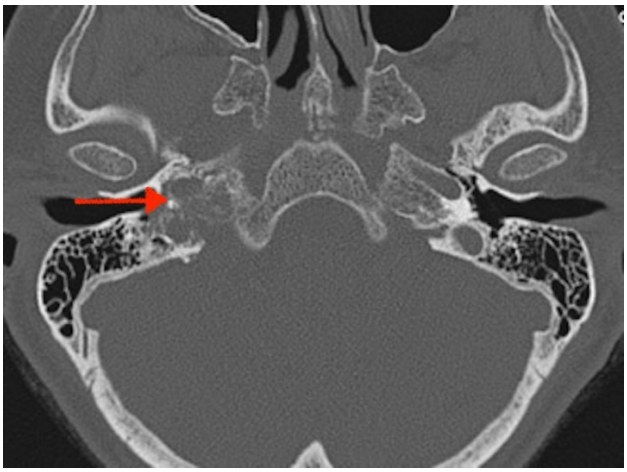

Figure 2. Axial CT temporal bone without contrast showing right glomus jugulare with classic irregular erosion and "moth eaten" appearance to temporal bone with extension to lower right CP angle and extending superolaterally towards hypotympanic region with erosive changes in the jugular bulb

Urine and plasma metanephrines were slightly elevated and she exhibited no symptoms of excess catecholamine. The metaiodobenzylguanidine scan was negative for pheochromocytoma.

2.2. Treatment

The patient was offered genetic testing but declined. She was started on low dose amitriptyline for neurogenic cough which significantly improved. Surgical management of the right extensive glomus jugulare was deferred as sacrifice of cranial nerves would be detrimental given the bilateral nature of the disease. She received 50.4 Gy external beam radiotherapy with 28 fractions to the right parapharyngeal space.

2.3. Outcome and Follow up

Patient tolerated the radiation therapy well. The patient reported moderate odynophagia and hypogeusia but denied dysphagia. At the conclusion of therapy, no skin reaction or mucositis was visible within the treatment fields. Patient was kept under close observation with clinical examination and imaging. She had symptomatic improvement in the tinnitus and cough had dramatically improved. However, she continued to have sensorineural hearing loss in her right ear.

Patient was followed up with serial imaging with contrast enhanced CT scans with a baseline scan at 3 months and then at 6 months interval for the first 2 years and then annually. At the last CT scan which was at 2 years after the completion of therapy, the paragangliomas were stable in size and no new lesions were identified.

3. Discussion

Paragangliomas are neuroendocrine tumors that arise from autonomic paraganglia. The majority arise in the head and neck from the glossopharyngeal and vagus nerves and can be associated with the carotid body, jugulotympanic, vagal, or even laryngeal paraganglia though the latter is rare. The majority of them are non-secretory and present with head and neck manifestations, such as neck masses, hoarseness, dysphagia, cough, or vertigo. [1] They can also arise anywhere outside of the head and neck along the sympathetic chain, with these much more likely to be secretory (86%). Catecholamine secreting paragangliomas can present with systemic effects of the catecholamines, which account for only about 5% of initial presentations. These systemic effects include sustained hypertension, headaches, palpitations, and perspiration, due to excessive production of norepinephrine and normetanephrine. [2,3]

About 30-50% are hereditary/syndromic, with those arising in head and neck typically associated with mutations in succinate dehydrogenase subunit (*SDH*) gene. [4,5,6,7] Other associated hereditary syndromes are MEN2, NF1, and vHL. [8] Multifocal tumors are more likely to occur in patients with *SDHD* gene mutations, while risk of malignancy is higher in patients who carry the *SDHB* germline mutation. Therefore, it is recommended that all patients diagnosed with paraganglioma undergo genetic testing. [4,5,7]

Malignant transformation is possible, but rare. Malignancy is defined by the presence of metastasis at non-chromaffin sites. There are no histopathologic or immunohistochemical features that indicate the possibility of malignant transformation. However, there are several clinical markers that may be able to predict metastatic potential, including younger age, large tumor, and special secretory profiles. [4,9,10]

Management, which may include surgery or radiation therapy, is complex and varies from case to case depending on symptomatology. Complete surgical excision of the tumor is favored when possible, but is associated with hemorrhagic, cerebrovascular, and neurological risks. [2,11,12]

This case highlights a rare presentation, workup, and treatment of several synchronous paragangliomas. To our knowledge, there is only one other case of five synchronous paragangliomas reported in the literature, in which a 38-year-old Thai male presented with bilateral carotid body tumors, bilateral jugulotympanic paraganglioma, and unilateral subclavian paraganglioma with no family history of paragangliomas. Prior to this, no incidence of more than three synchronous paragangliomas have been reported. [10]

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