Severe Dry Cough as Presenting Symptom of Patient with Five Paragangliomas

John Neiner MD, Vyas Rao MD, Mansour Mirfakhraee MD,
Maura Cosetti MD, Cherie-Ann Nathan MD
Department of Otolaryngology, LSU Health Shreveport

Abstract

Objectives: A case report of a 60 year old female who presented to a pulmonologist with dry cough and in the workup was found to have five paragangliomas – right glomus jugulare, two left glomus vagale, right glomus vagale, and left superior mediastinal paraganglioma.
Study Design: Case Report
Methods: Clinical history, radiology, and management of this patient are reviewed along with the current literature on multiple paragangliomas.
Results: The patient was referred to otolaryngology for workup and management. She was asymptomatic other than a cough with no other cranial nerve deficits. She did have longstanding right ear sensorineural hearing loss secondary to acoustic trauma. Masses were found to be non-secretory. Patient's cough resolved with amitriptyline and she elected to pursue external radiation for the most symptomatic right glomus jugulare.
Conclusion: This case highlights the presenting symptoms, multidisciplinary workup, and management of a rare case of five paragangliomas.

Introduction

Paragangliomas are neuroendocrine tumors that arise from autonomic paraganglia. The majority of them are non-secretory and present with head and neck manifestations. Catecholamine secreting paragangliomas can present with systemic effects of the catecholamines, which account for only about 5% of initial presentations. About 30-50% are hereditary/syndromic, with those arising in head and neck typically associated with mutations in SDH gene. Other associated hereditary syndromes are MEN2, NF1, VHL, and others. 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. They can also arise anywhere outside of the head and neck along the sympathetic chain, with these much more likely to be secretory (86%). Malignant transformation is possible, but rare.

Case Report

A 60 year old white female developed a dry cough of 8 month duration, treated initially with conservative management by PCP. A chest CT was performed which noted a contrast enhancing mass of the superior mediastinum/lower neck and was referred to ENT. She denied any dysphagia or voice changes. During the workup she began to note dull right ear pain. She had a history of a prior acoustic trauma with known deafness in her right ear for 25 years. On exam, she had a reddish mass at the inferior right tympanic membrane and a bilateral upper neck fullness/mass. Cranial nerves appeared intact. A CT neck and temporal bone was ordered as well as metanephrines. These scans showed five paragangliomas – a right glomus jugulare, 2 left glomus vagale, right glomus vagale, and a left superior mediastinum paraganglioma. Urine and plasma metanephrines were slightly elevated and she exhibited no symptoms of excess catecholamine. MIGB scan was negative for pheochromocytoma. Patient was offered genetic testing but declined. She was started on low dose amitriptyline for neurogenic cough which significantly improved. We discussed options with patient and she elected to pursue external beam radiation for her right glomus jugulare.

Radiology

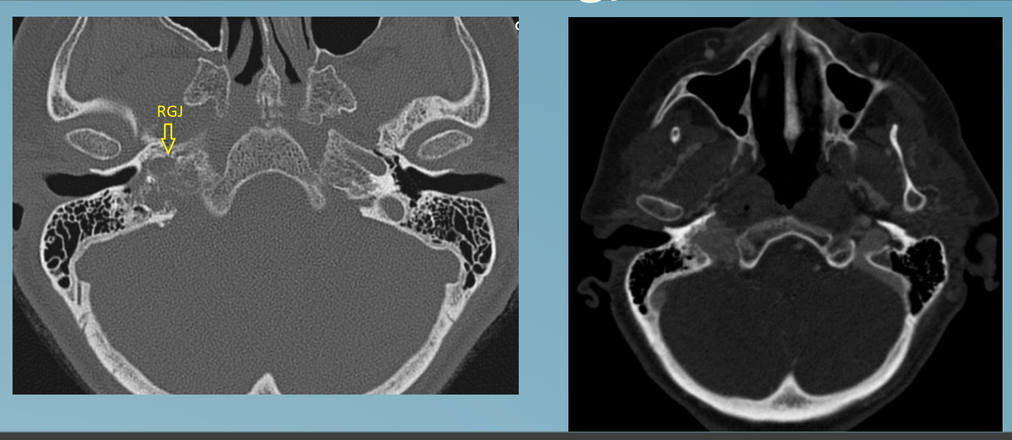

Figure 1: Axial CT temporal bone with contrast showing right glomus jugulare (RGJ) with classic irregular erosion and "moth eaten" appearance to temporal bone with extension to lower right CP angle and extending superolaterally towards hypotympanic region with erosive changes in jugular bulb.

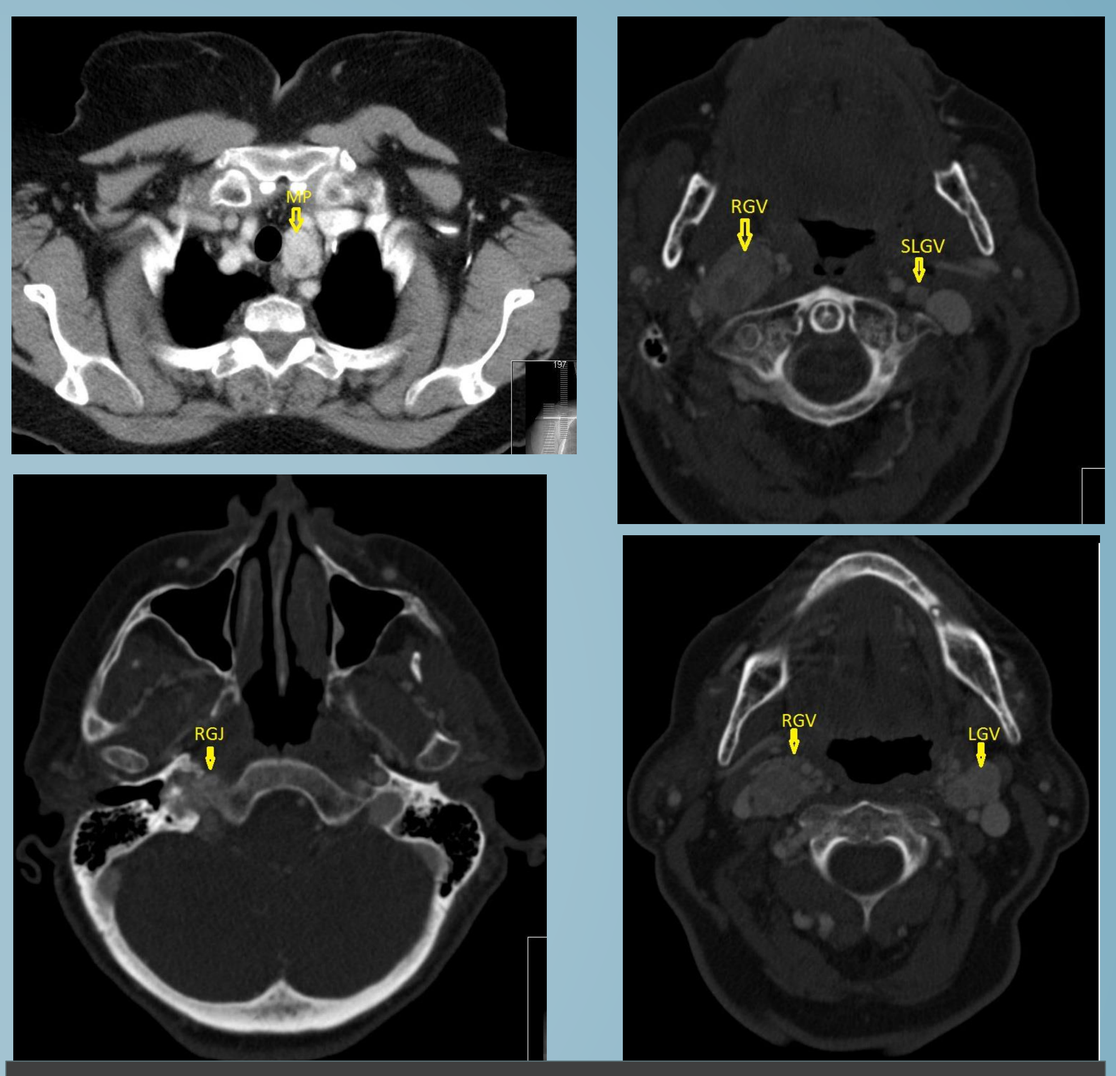

Figure 2: Axial CT neck and chest with contrast showing superior mediastinal paraganglioma (MP) between the proximal left common carotid and subclavian arteries, right glomus vagale (RGV) extending to a point just above carotid bifurcation with the carotid aretery displaced anteromedially, a lower left glomus vagale (LGV) and a smaller left glomus vagale (SLGV) high in the neck.

Discussion/Conclusion

In literature review we found one prior case report of five synchronous paragangliomas of the head and neck. This case highlights a rare presentation, workup, and treatment of several synchronous paragangliomas. The initial clinical manifestations of such can vary widely, such as this patient who presented with dry neurogenic cough presumably from vagal compression/involvement. The management is complex and varies from case to case depending on symptomatology. Our patient was managed with amitriptyline for her cough which provided symptomatic relief. Surgical management of the right glomus jugulare was deferred due to extension to cerebello-pontine angle just under internal auditory canal with intact CN. She was referred for external beam radiation to her right glomus jugulare for her worsening ear pain and headache as well as extension to cerebello-pontine angle and will be followed with serial exams and imaging.

Contact

Cherie-Ann Nathan M.D.
LSU Health Shreveport Otolaryngology
Email: cnatha@lsuhsc.edu
Website: http://myhsc.lsuhscshreveport.edu/OtolaryngologyHeadandNeckSurgery/ENTHomePage.aspx
Phone: 318-675-6262

References

1. Boedeker CC, Neumann HP, Maier W, et al. Malignant head and neck paragangliomas in SDHB mutation carriers. Otolaryngol Head Neck Surg 2007; 137:126.
2. Brown ML, Zayas GE, Abel MD, et al. Mediastinal paragangliomas: the mayo clinic experience. Ann Thorac Surg 2008; 86:946.
3. Drovdlic CM, Myers EN, Peters JA, et al. Proportion of heritable paraganglioma cases and associated clinical characteristics. Laryngoscope 2001; 111:1822.
4. Erickson D, Kudva YC, Ebersold MJ, et al. Benign paragangliomas: clinical presentation and treatment outcomes in 236 patients. J Clin Endocrinol Metab 2001; 86:5210.
5. Fishbein L, Merrill S, Fraker DL, et al. Inherited mutations in pheochromocytoma and paraganglioma: why all patients should be offered genetic testing. Ann Surg Oncol 2013; 20:1444.
6. Jackson CG, Glasscock ME 3rd, Harris PF. Glomus Tumors. Diagnosis, classification, and management of large lesions. Arch Otolaryngol 1982; 108:401.
7. Neumann HP, Pawlu C, Peczkowska M, et al. Distinct clinical features of paraganglioma syndromes associated with SDHB and SDHD gene mutations. JAMA 2004; 292:943.
8. Yavapolkul D, Jittreetat T. Five synchronous paragangliomas of the head and neck: A case report. J Med Assoc Thai 2016;99:S21-5.