Vagal Paraganglioma Associated With Cough on Palpation

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Abstract

Etiologic Objective: At the conclusion of this presentation, the participants should be able to recognize the typical clinical features of vagal paraganglioma, be aware vagal paraganglioma can be associated with cough, and understand pathogenesis and available management options.

Methods: Medical records for the patient were extensively reviewed. A MEDLINE search for key words "vagal paraganglioma" and "glossus vagalis" was performed, and abstracts were reviewed for cough as a symptom.

Results: A 70-year-old female presented with a neck mass associated with a dry cough triggered by palpation of the mass. Bouts of coughing were often of such severity that she would vomit. On examination, a neck mass deep to sternocleidomastoid was palpable which was discrete, mobile and non-palpable. Palpation of the mass immediately triggered coughing. Surgical excision of the mass was performed, with a final diagnosis of vagal paraganglioma.

Conclusions: To our knowledge, this is the first description of a vagal paraganglioma presenting as a neck mass associated with a severe cough. Furthermore, this uniquely could be triggered by palpation of the mass. This case is presented to aid physicians should they encounter a neck mass associated with cough. Vagal paraganglioma, although rare, should be part of the differential in such a presentation.

Case Report

A 70-year-old female presented with a 4-week history of a neck mass. She had suffered a concurrent sporadic dry cough unrelated to eating, talking, or any particular movement; however she reported that touching the swelling triggered fits of coughing often of such severity that she would vomit. She reported concomitant intermittent hoarseness. She denied dysphagia,odynophagia or any symptoms of velopharyngeal insufficiency. She had no family history of paragangliomas or other neuroendocrine tumors, nor any relevant past medical history. On examination, a 2.5 cm right level two neck mass deep to sternocleidomastoid was present, which was discrete, mobile and non-palpable. Palpation of the mass immediately triggered coughing. The remainder of the examination was unremarkable apart from a mildly hoarse voice and prominent retropharyngeal carotid arteries.

A blood panel was normal and 24-hour urine was negative for catecholamines. CT angiography of the neck demonstrated a heterogeneously enhancing irregular 2.4 x 2.2 x 2.2 cm mass within the right carotid sheath along the lateral aspect of the proximal internal and external carotid arteries with minimal spaying of these arteries (Figures 1 A, B, and C). The central area of the mass was of lower attenuation suggestive of necrosis. MRI displayed a 2.5 x 1.9 x 3 cm mass arising from the inferior margin of the right carotid to the level of the pharyngeal sinuses within the carotid sheath which intensely enhanced following contrast administration (Figures 2 and 3). The mass was intensely avid on octreotide scan which could be triggered by palpation of the mass.

The mass was excised through a 7 cm incision made 2 finger-breadths below the mandible, overlying the sternocleidomastoid. The mass was dissected free from the internal and external carotid arteries, and the internal jugular vein. The vagal nerve was intimately involved with the mass, thus required suture ligation and removal with the mass. The spinal accessory and hypoglossal nerves were identified and preserved. A right neck dissection and partial sternocleidomastoid neck excision was also performed. The pathology report described a completely excised encapsulated 3.5 x 2 x 1.5 cm tumor associated with a nerve segment and ganglion. The central 10% of the tumor showed tumor necrosis. Immunohistochemical staining for SDHB was positive, with intrathoracic heterogeneity, suggesting a germline mutation was not the cause for the tumor. All dissected lymph nodes were negative for malignancy.

Figure 1 A, B, and C. CT angiography of the neck with axial (A), coronal (B) and sagittal (C) sections of the heterogeneously enhancing irregular 2.4 x 2.2 x 2.2 cm mass within the right carotid sheath.

Discussion

Pathogenesis

• Paragangliomas arise from the paraganglia - small groups of neuroendocrine cells arising from autonomic nervous system ganglia.

• Usually slow growing and benign, tumors of the paraganglia are most common in the adrenal medulla (pheochromocytoma), with 85% of extra-adrenal PGs in the abdomen, 12% in the thorax, and 3% in the head and neck.

• Four genetic PG syndromes have been described, all with autosomal dominant transmission. Germline mutations are known in 3, all in the gene complex encoding succinate dehydrogenase (SDH): with subunits SDHD, SDHC or SDHB mutated, any of which predisposes PGs.

• More than half of head and neck PGs are sporadic.

Management

• Surgical excision is the historic treatment of choice for most VPs, however contemporary management is evolving toward more conservative measures due to the high associated morbidity.

• VP resection almost always requires vagus nerve sacrifice with resultant speech, swallow, and sensory deficits.

• Observation with serial imaging has been successfully employed, with the majority of lesions remaining radiologically stable, although neuropathy progression has occurred in a third of cases.

• Observation is especially valuable for asymptomatic older patients.

• Stereotactic radiosurgery is a proven treatment option for VPs <3cm in maximum dimension and should be offered as a potential first-line treatment.

Case in Context

To the best of our knowledge, this is the first description in the English language of a vagal paraganglioma presenting as a neck mass associated with a severe cough which could be triggered by palpation of the mass.

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References:


