



PULMONARY PUZZLES

A rare cause of chronic cough

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Received 1 August 2018

Revised 27 November 2018

Accepted 10 December 2018

Published Online First

12 January 2019

A 64-year-old female patient was referred to the pulmonary outpatient clinic with a 3-year history of non-productive cough and intermittent wheezing. The cough was mainly present during the day and could be triggered by talking or drinking cold drinks. She also mentioned seasonal rhinoconjunctivitis during spring and summertime. Clinical examination was unremarkable. Chest X-ray and pulmonary function tests were normal. A histamine provocation test was borderline positive (PD₂₀=0.37 mg). Laboratory analysis showed positive radioallergosorbent tests for grass and tree pollen. A diagnosis of cough-predominant, atopic asthma was made and inhaled corticosteroids (fluticasone) were prescribed. Three months later, the patient returned for a follow-up visit. Her cough had only partially improved. She now mentioned that her cough could also be triggered by rotating her head to the left or by touching the right side of her neck, where she occasionally had noticed a swelling of fluctuating dimensions. On clinical examination, no mass was palpable but the cough could indeed be elicited by touching the aforementioned region.

Contrast-enhanced CT angiography (figure 1A) and MRI (figure 1B) of the neck were performed, both revealing a 11×12×30 mm slight heterogeneously enhancing hypervascular mass, located in the right carotid space, between the common carotid artery and internal jugular vein, cranially extending along the lateroposterior side of the carotid bifurcation, without splaying of the internal and external carotid artery. The mass showed intense somatostatin receptor expression on Gallium-68 DOTATATE positron emission tomography-CT scan (figure 1C).

QUESTIONS

1. Which element(s) brought forward at the follow-up visit made us reconsider the initial diagnosis of cough-predominant asthma?
2. After examining the images provided, what is the most likely diagnosis?
3. How can this diagnosis explain the patient's cough?
4. What are the therapeutic options?

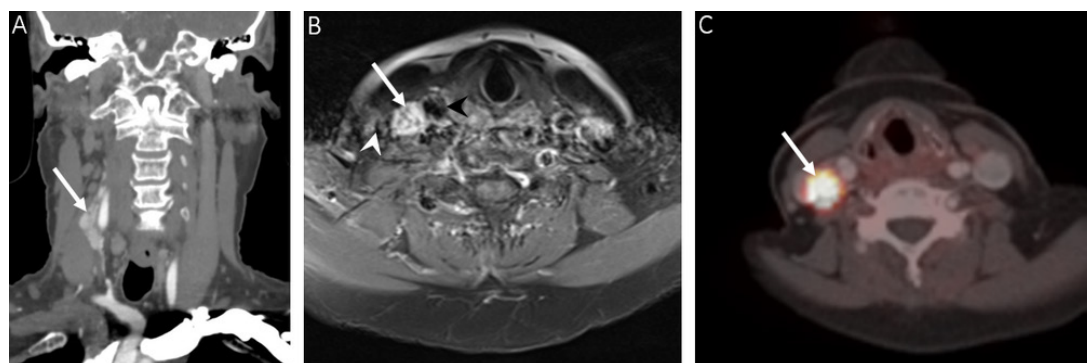


Figure 1 (A) Contrast-enhanced CT angiography, coronal reconstruction, showing an intensely enhancing mass (white arrow) along the distal part of the right common carotid artery. (B) MRI, axial image T1 weighted, with contrast and fat saturation, showing the same lesion (white arrow) between the right common carotid artery (black arrowhead) and internal jugular vein (white arrowhead). (C) Gallium-68 DOTATATE positron emission tomography (PET)-CT scan, fused axial section of Gallium-68 DOTATATE PET and contrast-enhanced CT, showing strongly increased uptake, representing somatostatin receptor expression, in the lesion (white arrow).



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To cite: Decoster L, Van der Goten A, Eerens I, *et al.* *Thorax* 2019;**74**:521–522.

ANSWERS

1. In this atopic patient with positive bronchial challenge, testing a diagnosis of cough-predominant asthma would have been very plausible. However, chronic cough guidelines suggest further evaluation in patients whose cough persists despite adequate treatment with inhaled corticosteroids.¹ The clear association between touching the neck and cough resulted in the decision to perform imaging of the neck as next diagnostic step.
2. The findings on imaging were compatible with a cervical paraganglioma. Paragangliomas are rare neuroendocrine tumours that arise from extra-adrenal autonomic paraganglia, small organs derived from the embryonic neural crest. Often asymptomatic and thus an incidental finding on imaging, paragangliomas may cause symptoms due to catecholamine hypersecretion or mass effect. Cough as the first presentation of a paraganglioma is rare.^{2,3}
3. Concerning the mechanism of cough, we hypothesise that the mass causes mechanical stimulation of small branches of the vagus nerve, similar to the Arnold's nerve ear-cough reflex, where manipulation of the external auditory canal can activate the auricular branch of the vagus nerve and evoke cough.⁴
4. Resection is the treatment of choice for secreting or locally symptomatic tumours. In non-catecholamine secreting paragangliomas, radiotherapy or stereotactic radiosurgery are safe and effective alternative treatment options, especially when resection would require sacrifice of critical vascular or neural structures. However, in small, pauci-symptomatic, non-secreting paragangliomas, an observational approach is acceptable.⁵ In the particular case of a paraganglioma causing cough, presumably due to sensory vagal neuropathy, a therapeutic trial with gabapentin or pregabalin can be considered. These neuromodulators significantly improved cough-specific quality of life measures in small randomised placebo-controlled trials in patients with chronic refractory

cough.⁶ Furthermore, Ryan *et al* described two patients with Arnold's nerve ear-cough reflex, successfully treated with gabapentin.⁴ Potential side effects and the risk-benefit profile of these drugs should be discussed and reassessment after 6 months of therapy is warranted.

In our patient, 24-hour urine collections were repeatedly negative for catecholamines and metanephrines consistent with a non-secreting paraganglioma. The different therapeutic options were discussed with the patient and a conservative approach with serial imaging was chosen. Three years after the initial diagnosis, the paraganglioma remains stable, without worsening of her symptoms. A therapeutic trial with gabapentin/pregabalin was not undertaken as impact of cough on quality of life was low (Leicester Cough Questionnaire total score 19).

Contributors LD drafted the manuscript. LD and ADR took part in the clinical management of the patient. AVdG, IE and KG were responsible for image acquisition. All authors approved the final version of the manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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